International Journal of Emergency Medicine

CASE REPORT Open Access

Unveiling three accessory spleens in one patient: a rare case report and literature review

Hamdah Hanifa[1], Hussein Alhussein[2*], Lissa Mahmandar[2], Sara Kadi[2], Mahfoud Najjar[3] and Ahmad Alhaj[4]

Abstract

Background During embryogenesis, the spleen undergoes intricate developmental processes, originating from mesenchymal cells in the dorsal mesentery. An accessory spleen, a common anomaly found in autopsies and abdominal CT scans, can often be mistaken for different types of tumors. To the best of our knowledge, this is the first case in Syria documenting the occurrence of 3 accessory spleens in a patient who had previously undergone splenectomy.

Case presentation A 33-year-old male presented with right hypochondrium pain, sharp and radiating to the right flank, exacerbated by movement and large meals. Past medical history included mild Irritable Bowel Syndrome (IBS) and splenectomy due to a traumatic accident in childhood. On admission, vital signs were stable, with abdominal tenderness in the right upper quadrant. Laboratory investigations showed normal values. Ultrasound revealed a lobulated mass at the right adrenal gland (4.5 × 5 cm) with an isoechoic to hypoechoic texture. Multi-slice computed tomography (MSCT) Scan showed multiple nodules in the right adrenal gland, regular in shape, exerting a compressive mass effect, and significant lymphadenopathy around the abdominal aorta. Elevated metanephrine levels raised suspicion of an extra-adrenal pheochromocytoma. Laparoscopic surgery was performed, revealing accessory spleens and normal adrenal tissue with no malignancy.

Conclusion Healthcare providers should consider accessory spleens as a differential diagnosis for masses near the adrenal glands. Multiple accessory spleens in the adrenal region can complicate cases. Accessory spleens in uncommon locations like the adrenal glands can be challenging to diagnose and manage.

Keywords Accessory spleen, Case report, Misdiagnosis, Splenosis, Pheochromocytoma, Adrenal tumour

Background

During embryonic development, the spleen, which originates from mesenchymal cells of the dorsal mesentery, goes through intricate processes [1]. An accessory spleen, a small piece of splenic tissue that forms abnormally during fetal development, is a common anomaly found in 10% to 30% of autopsies and seen in 16% of individuals undergoing abdominal CT scans with contrast dye [2]. Approximately 19% of humans have accessory spleens, which tend to enlarge after a splenectomy [3]. This accessory spleen, located in the retroperitoneal space, can sometimes be mistaken for tumors such as ganglion tumors, adrenal masses, or neuroendocrine

*Correspondence:
Hussein Alhussein
hussein2002net@gmail.com
[1] Faculty of Medicine, University of Kalamoon, Al-Nabk, Syria
[2] Faculty of Medicine, University of Aleppo, Aleppo, Syria
[3] Department of Surgery, General Surgery Division, Aleppo University Hospital, Aleppo, Syria
[4] Department of Surgery, Faculty of Medicine, University of Aleppo, Aleppo, Syria

tumors, particularly in hypertensive patients [4]. Retroperitoneal tumors, which can be benign or malignant, may include neurogenic tumors like pheochromocytoma, a rare tumor that produces catecholamines and originates from chromaffin cells of the adrenal medulla or paraganglion [5]. The clinical symptoms of pheochromocytoma, such as hypertension, palpitations, headaches, and excessive sweating, can vary and mimic other conditions [6]. This case report discusses a rare case of a 33-year-old patient who had a splenectomy during childhood and was initially misdiagnosed with a retroabdominal tumor due to an enlarged attached spleen. Further investigation revealed masses near the liver's navel within the peritoneum, putting pressure on the adrenal glands and raising suspicion of pheochromocytoma.

Case presentation

A 33-year-old male presented with a chief complaint of right hypochondrium pain, which began as episodic pain one year prior. The pain was described as sharp and radiated to the right flank, exacerbated by movement, lying on the right side, and consuming large meals. The patient denied any associated symptoms such as vomiting, diarrhea, nausea, or other gastrointestinal complaints. His past medical history was notable for mild Irritable Bowel Syndrome (IBS), with episodes typically triggered by the consumption of legumes and large quantities of bread. The patient had a surgical history of splenectomy at age seven following a traumatic accident. He was a daily waterpipe smoker for five years but had no history of alcohol use, drug use, allergies, or family history of significant medical conditions. His weight and appetite were within normal limits.

On admission, the patient's vital signs were as follows: pulse rate of 80 beats per minute, blood pressure of 130/80 mmHg, body temperature of 37.6 °C, and blood oxygen saturation (SpO2) of 98%. Clinical examination revealed a midline abdominal scar consistent with previous surgery, along with tenderness in the right upper quadrant and right flank, without rebound tenderness. The rest of the physical examination was unremarkable.

Laboratory investigations showed hemoglobin)Hgb(15.5 g/dL, white blood cell count) WBC(6.12×10^9/L, granulocytes percentage) Gran%(62.4%, platelet count) Plt(331×10^9/L, sodium)Na(137 mmol/L, potassium (K(4.3 mmol/L, Glucose 98 mg/dL, Urea 24 mg/dL, Creatinine 0.8 mg/dL, prothrombin time (PT) 13.7 s, activity 96%, and international normalized ratio (INR) 1.02 (see Table 1).

An ultrasound of the abdomen and pelvis was performed, which revealed lobulated mass at the anatomical location of the right adrenal gland, measuring 4.5×5 cm, with an isoechoic to hypoechoic texture. Doppler ultrasound was inconclusive for vascular assessment of the mass. Additionally, a similar 2 cm mass was identified near the porta hepatis, though its precise nature could not be determined. Several oval-shaped lymph nodes were detected, with the largest on the left side measuring 3×8.5 mm and on the right side measuring 6.5×16 mm. Lymph nodes with a fatty core were also observed around the femoral vessels, the largest on the right measuring 19×9 mm and on the left 12.5×7 mm. Importantly, there was no significant enlargement of lymph nodes around the iliac or axillary vessels.

Table 1 Laboratory tests

Investigations	At presentation	Normal range
Hemoglobin (Hgb)	15.5 g/dL	13.0-16.0 g/dL
White Blood Cells (WBC)	6.12×10^9/L	$4.5\text{-}13.0\times10^9$/L
Sodium (Na)	137 mmol/L	135-145 mmol/L
Potassium (K)	4.3 mmol/L	3.5-5.1 mmol/L
Glucose	98 mg/Dl	70-100 mg/dL
Urea	24 mg/Dl	7-20 mg/dL
Creatinine (Crea)	0.8 mg/Dl	0.6-1.2 mg/dL
Prothrombin Time (PT)	13.7 seconds	11-14 seconds
International Normalized Ratio (INR)	1.02	0.8-1.2

An overview of the laboratory values of the patient upon admission

Given these findings, the patient was prepared for further evaluation, a multi-slice computed tomography (MSCT) scan with contrast injection was performed, revealing multiple nodules in the right adrenal gland. These nodules were defined, regular in shape, and appeared to exert a compressive mass effect on the adrenal gland, possibly representing lymph node aggregation or metastasis. Additionally, significant lymphadenopathy was noted around the abdominal aorta (see Fig. 1).

Based on these imaging findings, the patient was referred to an endocrinologist before any surgical intervention. Urinary catecholamine metabolite levels were measured, showing metanephrine levels of 247.5 µg/24 h (normal range: < 350 µg/24 h) and significantly elevated normetanephrine levels of 1158 µg/24 h (normal range: < 600 µg/24 h), raising suspicion of an extra-adrenal pheochromocytoma. Other differential diagnoses included benign neoplastic lesion and adrenal cancer.

Consequently, the decision was made to proceed with laparoscopic surgery for biopsy and adrenalectomy, along with excision of the surrounding mass (see Supplementary Video 1). Informed written consent was obtained from the patient, who remained hemodynamically stable throughout the preoperative period without hypertensive episodes. The surgical procedure was uneventful, with intraoperative blood pressures averaging 110/80 mmHg, and no arrhythmias were observed. The resected

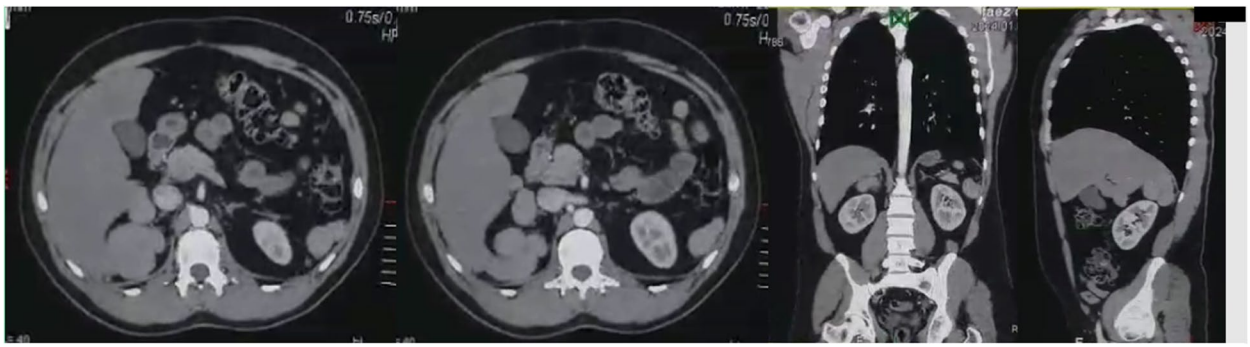

Fig. 1 Abdominal and Pelvic MsCT revealed tissue nodules in the anatomical area of the spleen. These nodules exhibit clear equivalent density, regular borders, and overlapping. The largest nodule measures 3.5 cm and appears to represent a lymph node as an additional splenic differential diagnosis. Additionally, on the right adrenal, several tissue nodules are tangential to the lateral peduncle, isolated from the medial peduncle, exerting a compressive mass effect. Lymphadenopathy is also observed around the abdominal aorta

specimens were sent for histopathological examination, which revealed the presence of three accessory spleens exhibiting signs of congestion. Additionally, the excisional biopsy of the abdominal mass confirmed normal right adrenal gland tissue, which was surrounded by abundant adipose tissue. A reactive lymph node was also identified, with no evidence of malignancy (see Fig. 2).

Postoperatively, the patient was monitored in the intensive care unit and remained stable, with the chest tube being removed prior to discharge. During a week-long hospital stay, the patient exhibited stable vital signs without any notable complications. The final diagnosis was three accessory spleens. He was discharged after one week with follow-up scheduled.

Discussion

The spleen is one of the important and essential organs for the immune system, as it represents the largest organ in the lymphatic system. It consists of white pulp, which contains lymphocytes, and red pulp, which gets its color from containing blood [7]. The importance of the spleen is manifested in its immune and defensive function in the human body, such as producing white blood cells, filtering the blood, and getting rid of microbes. Some individuals may experience certain congenital or acquired abnormalities in the spleen, including wandering spleen, accessory spleen, and polysplenia syndrome, in addition to abnormalities related to its shape [8]. Wandering spleen is a rare condition characterized by the complete

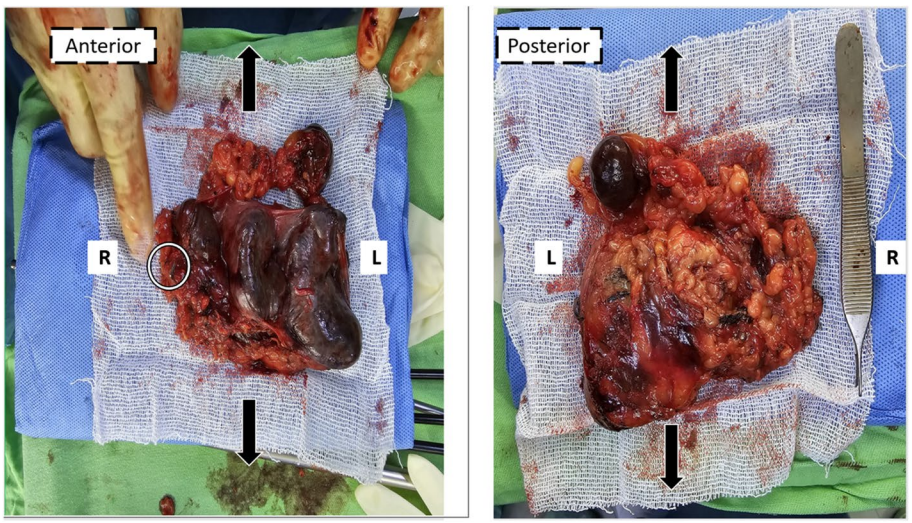

Fig. 2 Anterior and posterior views of the resected specimen showing multiple masses excised en bloc from the peritoneum. The anterior view displays masses identified as accessory spleens located in front of the kidney, while the posterior view highlights the excised adrenal gland along with surrounding adipose tissue, attached to the peritoneum

absence of splenic ligaments or failure of the spleen to remain in its normal position [9]. Wandering spleen has been reported in a few cases documented in the medical literature (around 500 cases) [10]. Hanifa et al. suggested a possible relationship between wandering spleen and lymphoma [11]. Polysplenia syndrome (PSS) is associated with multiple abnormalities in the body such as heart, lung, and pancreatic abnormalities, and is often discovered incidentally [12]. Although the syndrome primarily occurs in children, it has been frequently documented in adults in the medical literature [13]. El Mountassir et al. reported a case of PSS in a 54-year-old adult patient who had chronic abdominal pain, with the diagnosis later confirmed through computed tomography imaging [14]. Many abnormalities may be observed in the shape of the spleen, especially during autopsies, the most prominent of which is the persistence of the fetal splenic lobes beyond fetal life, which are located on the medial side of the spleen in most cases [15]. A lobulated spleen was discovered by medical students while studying anatomy on one of the cadavers. The appearance of the spleen consists of 7 lobes/lobules in addition to five fissures [16]. Celiac disease and sickle cell anemia are among the most important causes of the development of hyposplenia [17]. Patients with hyposplenia are susceptible to infection with various types of life-threatening bacteria, so vaccination and preventive antibiotics are important for their life safety. In one study, the first case documented in the medical literature was in a patient with hyposplenia who experienced a recurrence of invasive pneumococcal disease despite vaccination against pneumococcal disease [18]. Regarding splenosis, it is a condition that is not serious and occurs as a result of splenic rupture following surgery or trauma [19]. A case of thoracic splenosis was reported, in which autotransplantation of splenic tissue occurred in the left pleural space in a patient who suffered from chronic chest pain [20]. A case of failure of fusion of the splenial primitive buds that derive from the dorsal mesentery by the fifth week of embryogenesis is known as the accessory spleen (AS) [21]. It has been observed to occur in females at a greater rate than in males, with a peak incidence between 20–40 years (see Table 2) [22]. In our case, the patient was a male, 33 years old. AS may occur in several sites, most notably the tail of the pancreas, splenic ligament, greater omentum, and splenic hilum [23]. The presence of an accessory spleen in the scrotal sac is considered a unique condition, with the first documented case in medical literature by Sneath in 1913. In a previous study, two cases of accessory spleen in the scrotal sac were reported, one of which presented with pain in the inguinal region with a mass in the testicle, and the other with an empty bilateral scrotum with hypospadias since birth [24]. Additionally, an accessory spleen in the form of a pelvic mass was discovered behind the uterus in a 39-year-old woman who was suffering from severe abdominal pain. Although the presence of an accessory spleen in the pelvis is rare, it requires consideration as a differential diagnosis by gynaecologists [25]. Trujillo et al. reported on the accessory spleen in a patient suffering from iron-deficiency anemia, which appeared as a submucosal gastric lesion [26]. In our case, the patient had undergone splenectomy as a child, and after 26 years presented to the emergency room with complaints of pain in the right hypochondrium. He had 3 accessory spleens that appeared as tissue nodules spread at the mesentery, near the liver, and around the kidneys.

Table 2 Summary of the main points and prominent results from the literature

Condition	Description	Key Characteristics	Notable Findings
Wandering Spleen	Absence of splenic ligaments or failure to remain in normal position	Rare condition, around 500 cases documented	Possible relationship with lymphoma (Hanifa et al.)
Polysplenia Syndrome (PSS)	Multiple abnormalities in the body, often discovered incidentally	Associated with heart, lung, and pancreatic abnormalities	Confirmed via CT imaging (El Mountassir et al.)
Lobulated Spleen	Persistence of fetal splenic lobes beyond fetal life	Appearance consists of lobes/lobules and fissures	Discovered during autopsies (Nayak et al.)
Hyposplenia	Reduced spleen function, leading to susceptibility to infections	Susceptibility to life-threatening bacterial infections	First documented case of recurrent invasive pneumococcal disease despite vaccination (Ballegaard et al.)
Splenosis	Autotransplantation of splenic tissue following rupture	Occurs as a result of splenic rupture following surgery or trauma	Case of thoracic splenosis with splenic tissue in the left pleural space (Le Bars et al.)
Accessory Spleen (AS)	Small piece of splenic tissue formed during fetal development, often in abnormal locations	Observed more in females; peak incidence between 20-40 years	First documented in scrotal sac by Sneath in 1913 and in this case, we present the first reported case from Syria of a patient with three accessory spleens.

The various splenic abnormalities that were reviewed in this current study

As a result of the increase in normetanephrine and the presence of a large mass in the right adrenal gland, a pheochromocytoma was suspected, which led to the removal of the right adrenal gland. It was later revealed by the histopathological examination that the adrenal glands were normal and the masses were accessory spleens. In a similar previous case to ours, the patient had also undergone splenectomy in the past and was admitted at the age of 55 to the emergency department due to chest and abdominal pain. After further investigations and due to a history of high blood pressure and the presence of a mass in the left adrenal gland, the surgeons decided to completely remove the gland area, only to later discover that the excised mass was actually an accessory spleen [27]. This highlights the importance of considering an accessory spleen in the differential diagnosis when suspecting a mass near the adrenal glands to avoid misdiagnosis. The accessory spleen may be single in 85% of cases; Two accessory spleens may be present in 14% of cases; The presence of 3 or more accessory spleens is rare and constitutes 1%, which makes our case unique and rare due to the presence of 3 accessory spleens in our patient [28]. Accessory spleens are often discovered incidentally as they usually do not cause any symptoms. However, when they are found in uncommon locations such as the adrenal gland, as in our case, they can be clinically significant, be incidental, and may be mistaken for malignant tumors [29]. It is difficult to diagnose an accessory spleen before surgery. Nuclear medicine imaging is the only method that can definitively confirm a diagnosis, using scintigraphy with 99mTC-labelled colloids or TC-99 m heat damaged red blood cells [30]. In some cases the utility of diagnosing the accessory spleen via endoscopic ultrasound-guided fine needle aspiration has been demonstrated [26]. Regarding treatment, surgery is essential in cases of torsion of the accessory spleen and cases in which the accessory spleen causes complications such as bleeding, obstruction, and chronic pain. However, in the majority of cases, no treatment is needed for the AS [31].

Conclusion

In light of this case, it is crucial for healthcare providers to be vigilant in considering accessory spleens as a potential differential diagnosis when evaluating masses near the adrenal glands. The identification of multiple accessory spleens in our patient's adrenal region serves as a reminder of the unique and complex nature of such cases. Although accessory spleens are often benign and asymptomatic, their presence in uncommon locations like the adrenal glands can pose challenges in terms of diagnosis and management. Moving forward, research efforts should be directed towards better understanding the prevalence and clinical implications of accessory spleens in atypical sites, as well as developing optimal strategies for their detection and treatment.

Abbreviations

IBS	Irritable bowel syndrome
SpO2	Oxygen saturation
PSS	Polysplenia syndrome
AS	Accessory spleen
MSCT	Multi-slice computed tomography

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s12245-024-00758-3.

Supplementary Material 1.

Acknowledgements

Not applicable.

Authors' contributions

HH, HA, LM, SK, MN, have participated in writing the manuscript, and reviewing the literature. Dr. Hamdah Hanifa critically and linguistically revised the manuscript. Dr. Hamdah Hanifa and Dr. Ahmad Alhaj conceived and supervised the conduct of the study. All authors read and approved the final manuscript.

Funding

No funding was required in the preparation of the manuscript.

Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Ethical clearance was obtained from the ethical committee of Aleppo University Hospital and consent was obtained from our patient to prepare the case for case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this study and accompanying images.

Competing interests

The authors declare no competing interests.

Received: 16 September 2024 Accepted: 7 November 2024
Published: 12 November 2024

References

1. Li Y, Qiu X, Li W, Guo H, Li X. Diagnostic challenge for giant left retroperitoneal accessory spleen: a case report. J Int Med Res. 2020Feb;48(2):300060519875898.
2. Maharaj R, Ramcharan W, Maharaj P, Greaves W, Warner WA. Right sided spleen laying retro-duodenal: A case report and review of the literature. Int J Surg Case Rep. 2016;24:37–42.
3. ARBER DA. CHAPTER 42 - Spleen. In: Weidner N, Cote RJ, Suster S, Weiss LMBTMSP (Second E, editors. Philadelphia: W.B. Saunders; 2009. p. 1512–35. Available from: https://www.sciencedirect.com/science/article/pii/B9781416039662000424.
4. Zang G, Dong B, Zhu G, Qiu X, Zhao Y. Accessory spleen after splenectomy mimicking adrenal tumor: a case report. Transl Cancer Res. 2020;9:5679–83.

5. Liu Z, Ma J, Jimenez C, Zhang M. Pheochromocytoma: A Clinicopathologic and Molecular Study of 390 Cases From a Single Center. Am J Surg Pathol. 2021Sep;45(9):1155–65.
6. Reisch N, Peczkowska M, Januszewicz A, Neumann HPH. Pheochromocytoma: presentation, diagnosis and treatment. J Hypertens. 2006;24(12). Available from: https://journals.lww.com/jhypertension/fulltext/2006/12000/pheochromocytoma__presentation,_diagnosis_and.1.aspx.
7. Mebius RE, Kraal G. Structure and function of the spleen. Nat Rev Immunol. 2005;5(8):606–16. Available from: https://doi.org/10.1038/nri1669.
8. Kapila V, Wehrle CJ, Tuma F. Physiology, Spleen. In Treasure Island (FL); 2024.
9. Reisner DC, Burgan CM. Wandering Spleen: An Overview. Curr Probl Diagn Radiol. 2018;47(1):68–70.
10. Koliakos E, Papazarkadas X, Sleiman MJ, Rotas I, Christodoulou M. Wandering Spleen Volvulus: A Case Report and Literature Review of This Diagnostic Challenge. Am J Case Rep. 2020Sep;21: e925301.
11. Hanifa H, Al-Shami K, Al-Shaher T, Ataya J, Al-Abrass M, Moezzen H, et al. Gastric volvulus with necrosis and gangrene associated with wandering spleen: A rare case report from Syria. SAGE Open Med Case Rep. 2024;12:2050313X241262141.
12. Huseynova RA, Bin Mahmoud LA, Abdelrahim A, Alroiedy MA, Huseynov O. Polysplenia syndrome with complex heart disease and jejunal atresia with malrotation in neonate: A case report. Clin Case Rep. 2020;8(5):848–51. https://doi.org/10.1002/ccr3.2768.
13. Malki MC, Outznit M, Mechhor S, Bouibaouen B, Nkurunziza L, El Bacha H, et al. Polysplenia syndrome in adulthood: a case report. Pan Afr Med J. 2022;41:67 Uganda.
14. El Mountassir M, Borahma M, Benelbarhdadi I, Lagdali N, Hosni A, Ajana FZ. Polysplenia syndrome revealed in adulthood by pancreatic and vascular malformations: a case report. Pan Afr Med J. 2022;43:77 Uganda.
15. Gayer G, Hertz M, Strauss S, Zissin R. Congenital Anomalies of the Spleen. Semin Ultrasound, CT MRI. 2006;27(5):358–69. Available from: https://www.sciencedirect.com/science/article/pii/S0887217106000540.
16. Nayak SB, Shetty P, R D, Sirasanagandla SR, Shetty SD. A lobulated spleen with multiple fissures and hila. J Clin Diagn Res. 2014;8:AD01–2.
17. William BM, Corazza GR. Hyposplenism: A comprehensive review. Part I: Basic concepts and causes. Hematology. 2007;12(1):1–13. Available from: https://doi.org/10.1080/10245330600938422.
18. Ballegaard VC, Schejbel L, Hoffmann S, Kantsø B, Fischer CP. Recurrent severe invasive pneumococcal disease in an adult with previously unknown hyposplenia. BMC Infect Dis. 2015Apr;15:171.
19. Fremont RD, Rice TW. Splenosis: a review. South Med J. 2007Jun;100(6):589–93.
20. Le Bars F, Pascot R, Ricordel C, Corbineau H, Verhoye JP, Richard De Latour B, et al. Thoracic splenosis: Case report of a symptomatic case. Chin J Traumatol; 2020;23:185–6.
21. Yang B, Valluru B, Guo YR, Cui C, Zhang P, Duan W. Significance of imaging findings in the diagnosis of heterotopic spleen-an intrapancreatic accessory spleen (IPAS): Case report. Medicine (Baltimore). 2017Dec;96(52): e9040.
22. Smith HC, Kakar N, Shadid AM. Accessory Spleen Masquerading as an Intrapancreatic Tumor: A Case Report. Cureus. 2022;14:e24677.
23. Li BQ, Xu XQ, Guo JC. Intrapancreatic accessory spleen: a diagnostic dilemma. HPB. 2018;20(11):1004–11. https://doi.org/10.1016/j.hpb.2018.04.004.
24. Marwah N, Bhutani N, Kalra R, Kajal P. Journal of Pediatric Surgery Case Reports Accessory spleen within the scrotum. J Pediatr Surg Case Rep. November 2018;2019(42):19–23. https://doi.org/10.1016/j.epsc.2018.12.009.
25. Feng Y, Shi Y, Wang B, Li J, Ma D, Wang S, et al. Multiple pelvic accessory spleen: Rare case report with review of literature. Exp Ther Med. 2018Apr;15(4):4001–4.
26. Trujillo SG, Saleh S, Burkholder R, Shibli F, Shah B. Accessory Spleen: A rare and incidental finding in the stomach wall. Cureus. 2022;14:e24977.
27. Zou Y, Xie X, Yan S, Wu G, Liu Q. Case report: Misdiagnosis of accessory spleen in the left adrenal region as an adrenal tumour after splenectomy. Front Surg. 2022;9:1017603.
28. Palumbo V, Mannino M, Teodoro M, Menconi G, Schembari E, Corsale G, et al. An extremely rare case of an oversized accessory spleen: case report and review of the literature. BMC Surg. 2019;19(1):45. https://doi.org/10.1186/s12893-019-0510-z.
29. Yildiz AE, Ariyurek MO, Karcaaltincaba M. Splenic anomalies of shape, size, and location: pictorial essay. ScientificWorldJournal. 2013;2013: 321810.
30. Bajwa SA, Kasi A. Anatomy, Abdomen and Pelvis: Accessory Spleen. In Treasure Island;2024.
31. Kuroiwa M, Takayama H, Uchikawa Y, Shimada R. Surgical resection for accessory spleen torsion: A case report. Int J Surg Case Rep. 2023;102:107835. Available from: https://www.sciencedirect.com/science/article/pii/S2210261222010811.

Publisher's Note