



Agenesis Of Appendix: A Rare Case Report

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Abstract

Agenesis of vermiciform is a exceptionally rare congenital anomaly, most often identified incidentally during surgical exploration for suspected acute appendicitis. Its clinical significance lies in its ability to closely mimic acute appendicitis, frequently leading to diagnostic and intraoperative dilemmas. This case report discusses a 29 year old female who presented with acute onset abdominal pain and vomiting for last 24 hours, clinical examination revealed severe colicky pain localized to right iliac region suggestive of acute appendicitis. Ultrasound of the same day reported a mildly inflamed appendix with diameter 6.6 mm, enlarged surrounding lymph nodes and no abnormality related to adnexa, reinforcing the provisional diagnosis. Intraoperatively, the appendix was not found at the converge of taenia coli. Further exploration of right adnexa reveled a ruptured, twisted, hemorrhagic ovarian cyst on the right side, for which right oophorectomy was performed. Postoperative management included broad spectrum antibiotics, aseptic wound care and radiological follow ups, leading to significant clinical improvement.

The absence of the appendix obliges the surgeon to actively search for alternative causes of right iliac fossa pain, particularly gynecological conditions in females of reproductive age. Unnecessary caecal dissection may increase operative time and risk of complications such as serosal injury or postoperative ileus. When appendicular agenesis is confirmed and the underlying pathology is appropriately addressed, the prognosis is excellent, with no long-term adverse outcomes related to the anomaly itself. Awareness of this rare entity, combined with a systematic intraoperative approach and judicious decision-making, is crucial to prevent avoidable morbidity and to ensure optimal surgical management in patients presenting with a clinical picture suggestive of acute appendicitis.

Keywords: Agenesis of appendix, Appendicitis, Torsion of ovary, Caecal diverticulum, Appendiceal anomaly, rare case report

INTRODUCTION

Right iliac fossa pain is one of the most frequent presentations in emergency surgical practice, with acute appendicitis being the leading provisional diagnosis in young adults. Despite improvements in diagnostic algorithms and widespread use of ultrasonography and computed tomography, a definitive preoperative diagnosis is not always achievable, particularly in the presence of rare anatomical anomalies or coexisting

intra-abdominal pathology^{1,2}. Such situations may result in unexpected intraoperative findings that challenge standard surgical decision-making.

Agenesis of the veriform appendix is an exceptionally uncommon congenital condition, reported in less than 1 per 100,000 laparotomies performed for suspected appendicitis³. Embryologically, it is attributed to failure of differentiation or arrested development of the caecal diverticulum during the fifth to eighth week of gestation⁴. The anomaly has no specific clinical features and almost invariably presents with symptoms indistinguishable from acute appendicitis, making preoperative identification virtually impossible⁵.

The absence of the appendix at surgery creates a diagnostic dilemma and necessitates a structured and exhaustive exploration to rule out ectopic appendix, intramural appendix, or complete auto-amputation following intrauterine or postnatal inflammation⁶. Additionally, alternative causes of right iliac fossa pain such as caecal diverticulitis, mesenteric adenitis, Meckel's diverticulum, and gynecological emergencies must be actively considered, especially in females of reproductive age⁷.

Though appendicitis is commonly encountered surgical department, but agenesis of appendix mimicking acute appendicitis is rare. In this article the case of 29 year old female who diagnosed as appendicitis and operated right oophorectomy for intraoperatively revealing absence of appendix at expected site and incidental finding of ruptured, twisted and hemorrhagic cyst of right ovary has been described. The patients clinical condition improved with further treatment and local wound care, she was subsequently discharged.

CASE REPORT-

Patient information-

A 29-year-old female presented to the surgery department with a chief complaint of pain in the right lower abdomen for the past 24 hours. The pain was acute in onset and was associated with two episodes of vomiting during the same period. There was no history of fever, altered bowel habits, urinary symptoms, or similar episodes in the past. The patient had no known medical comorbidities and was not on any regular medication. There was no significant past medical history. Past surgical history was significant for a lower segment caesarean section (LSCS) performed four years ago. There was no history suggestive of gynecological disorders or previous abdominal inflammatory conditions. Laboratory investigations revealed an elevated leucocyte count rest all hematological markers were normal.

Clinical examination

Per Abdomen: On inspection, the abdomen appeared normal in contour with no visible distension or dilated veins. On palpation, there was localized tenderness in the right iliac fossa with guarding. Rebound tenderness was present. All classical clinical signs suggestive of acute appendicitis, including McBurney's point tenderness, psoas sign, and obturator sign, were positive. Rovsing's sign was negative. There was no palpable mass. Percussion revealed localized tenderness in the right lower quadrant. On auscultation, bowel sounds were present.

Per Rectal Examination: Per rectal examination revealed no external abnormalities. There was no anal spasm. The rectum was loaded with fecal matter. No tenderness, mass, or abnormality was appreciated on digital rectal examination.

Diagnostic assessment

Acute appendicitis was suspected based on the clinical presentation and positive appendicular signs. The patient had undergone an ultrasonography outside prior to presentation. The USG report showed Grade I fatty liver. In the right iliac fossa, the appendix was visualized and appeared mildly inflamed, measuring 6.6 mm in diameter with a wall thickness of 2.4 mm, along with surrounding mesenteric inflammation. These findings were suggestive of a mildly inflamed appendix, correlating with the clinical features.

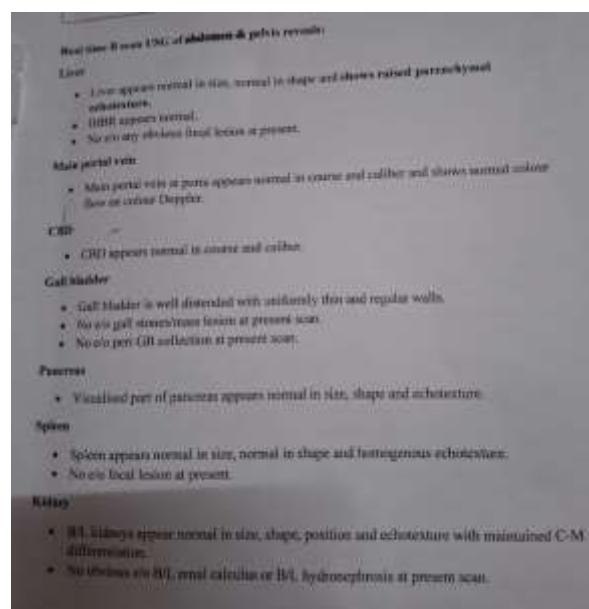


Fig (1)

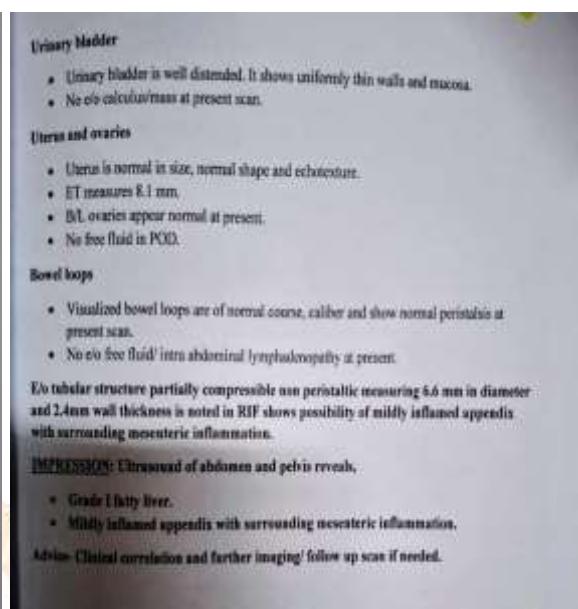


Fig (2)

Figure 1, 2 – Ultrasound of patient reporting mildly inflamed appendix

Surgical intervention

In view of the clinical and radiological features suggestive of acute appendicitis, the patient was taken up for appendectomy under spinal anesthesia through a McBurney's gridiron incision. On exploration, the caecum was identified and the taeniae coli were traced meticulously; however, the veriform appendix was not visualized at its usual anatomical location. To exclude an ectopic or abnormally positioned appendix, thorough caecal mobilization was performed and the visceral peritoneum was meticulously dissected. All possible locations of the appendix, including retrocecal, pelvic, subcecal, preileal, and postileal positions, were carefully explored. Despite exhaustive exploration, no appendicular structure could be identified, thereby raising a strong intraoperative suspicion of agenesis of the veriform appendix.

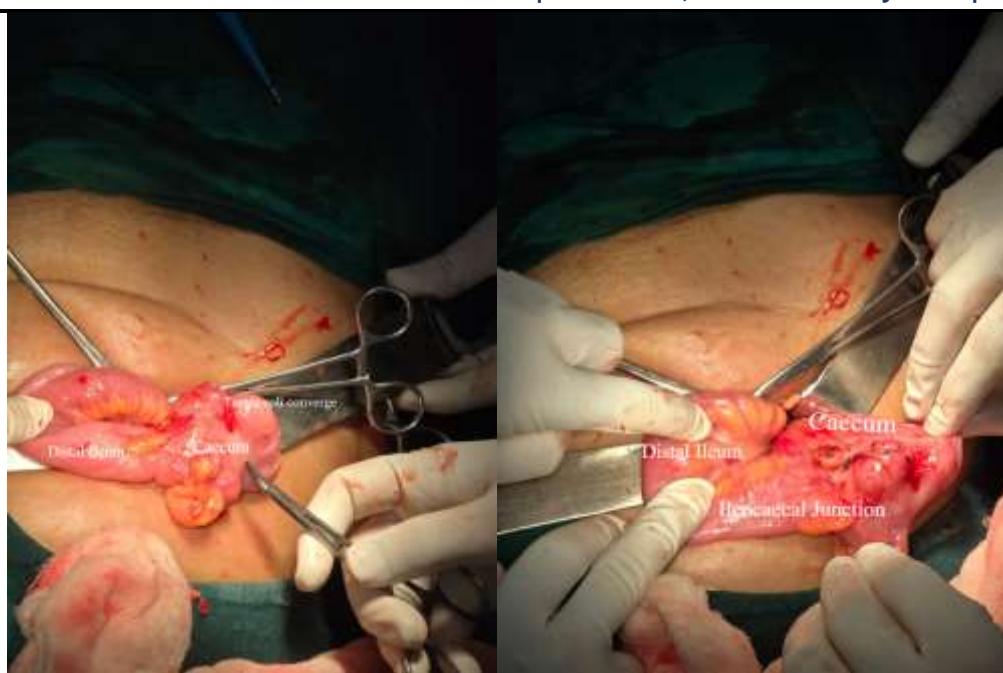


Fig (3)

Fig (4)

Figure 3 – Intraoperative picture of converge of taenia coli

Figure 4 – Dissected visceral peritoneum

Subsequent pelvic and adnexal examination revealed a right-sided twisted hemorrhagic ovarian cyst. The left ovary and uterus were normal in appearance. As the right ovary was non-viable, a right oophorectomy was performed. Adequate hemostasis was achieved, the operative field was irrigated, and the abdomen was closed in layers.



Fig (5)

Figure 5 – Adnexal examination showing right twisted, hemorrhagic cyst and left appears normal in physiology

Postoperatively, the patient showed significant clinical improvement and was managed with broad-spectrum antibiotics, analgesics, and local aseptic wound care. A postoperative contrast-enhanced CT scan of the abdomen and pelvis confirmed the absence of the veriform appendix, postoperative changes in the

right iliac fossa, and findings consistent with the treated right adnexal pathology, with no evidence of residual intra-abdominal collection, supporting the intraoperative suspicion of appendiceal agenesis. Also sample of right ovary send for HPE reported ovarian parenchyma with areas of hemorrhage.

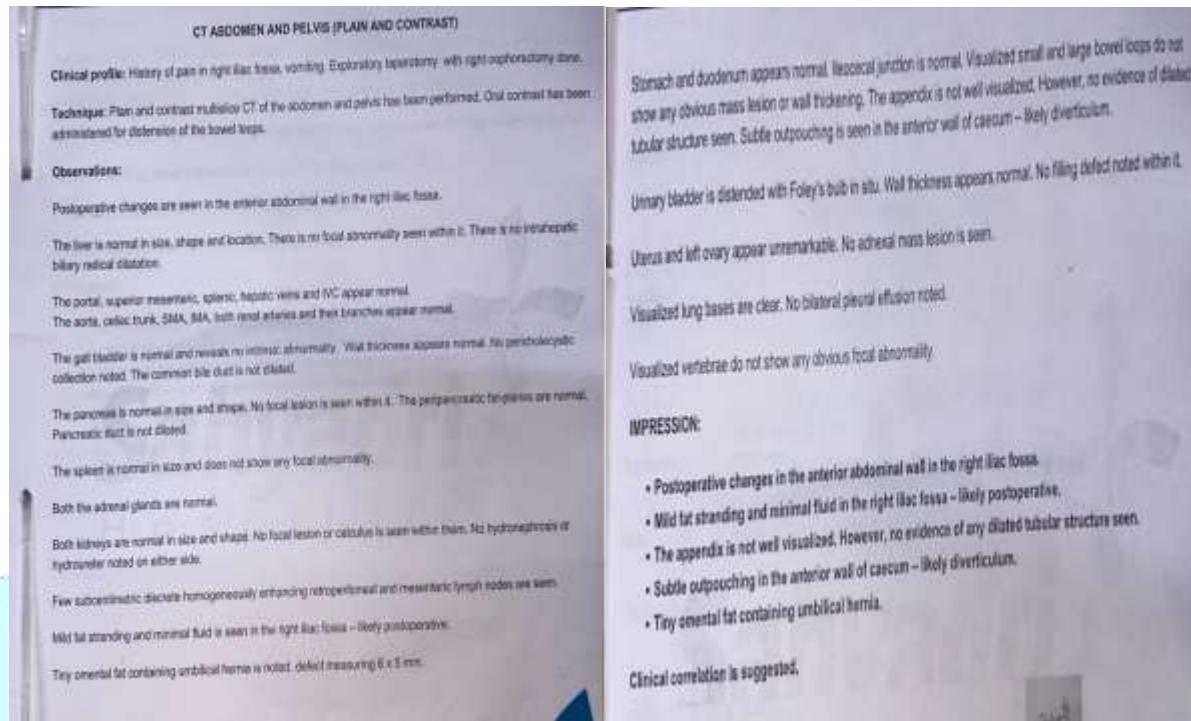


Fig (6)

Figure 6 – Post-operative CT abdomen and pelvis with contrast

Figure 7 – HPE report for right twisted, hemorrhagic cyst

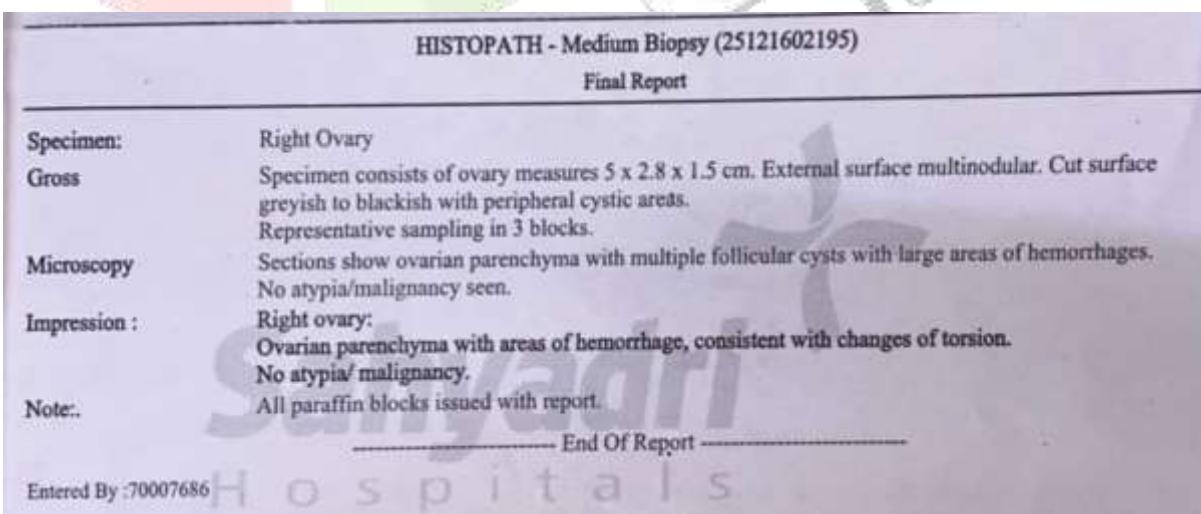


Fig (7)

Discussion

Agenesis of the veriform appendix is an extremely rare congenital anomaly and is most often identified incidentally during surgical exploration performed for suspected acute appendicitis. The present case highlights the diagnostic uncertainty that may arise even in the presence of classical clinical features and

supportive radiological findings and emphasizes the importance of meticulous intraoperative exploration, particularly in female patients presenting with right iliac fossa pain⁸.

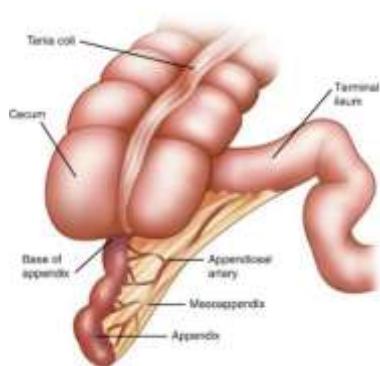


Fig (8)

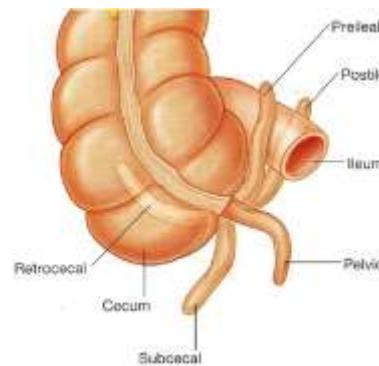


Fig (9)

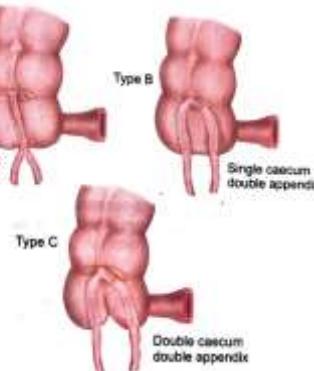
Fig (10)¹²

Figure 8 – Normal anatomy of Ileo-Caecal junction

Figure 9 – Multiple positions of appendix

Figure 10 – Congenital anomalies of appendix

The vermiform appendix is a narrow, blind-ended tubular structure arising from the posteromedial wall of the caecum, typically 2–3 cm below the ileocaecal junction at the convergence of the three taeniae coli. It consists of a base, body, and apex and contains abundant lymphoid tissue within its submucosa. Anatomical variations in the location of the tip of the appendix are common and clinically significant; retrocecal, pelvic, subcaecal, pre-ileal, post-ileal, and even subhepatic positions have been described, with retrocecal and pelvic positions being the most frequent⁹. These variations influence clinical presentation and may mimic other intra-abdominal or pelvic pathologies such as ureteric colic, Crohn's disease, or ovarian torsion.

Congenital anomalies of the appendix include duplication, hypoplasia, diverticula, and complete agenesis. Among these, appendiceal agenesis—defined as complete absence of the structure—is the rarest. The earliest description of appendiceal agenesis dates back to Morgagni in 1719, yet even nearly three centuries later it remains a seldom reported entity¹⁰. Based on autopsy and laparotomy series, the estimated incidence of appendiceal agenesis ranges from approximately 1 in 100,000 laparotomies performed for suspected appendicitis to as rare as 0.0009% of abdominal surgeries in historical series^{10,11}. Reports also indicate that agenesis may be identified in approximately 0.006% of autopsies, suggesting the anomaly exists sub clinically and is only detected when directly examined¹³.

From a modern embryological perspective, the appendix begins to form during the eighth week of gestation as a diverticulum of the caecum, which itself arises from the post-arterial segment of the midgut loop during physiological herniation and rotation. Normal appendicular morphogenesis depends on coordinated epithelial proliferation, mesenchymal differentiation, and adequate vascular supply. Disruption of these processes—whether due to localized developmental arrest, vascular insult, or genetic factors—may result in complete failure of appendicular development, yielding congenital agenesis^{8,14}. Agenesis may occur in isolation or, rarely, in association with other gastrointestinal anomalies such as ileal duplications, as described in pediatric cases¹⁵.

Clinically, acute appendicitis is one of the most common causes of acute abdomen, with a lifetime risk reported to be approximately 7–8% in the general population. Diagnostic tools such as the Alvarado score and imaging modalities—ultrasonography and contrast-enhanced computed tomography (CT)—are widely used to increase preoperative diagnostic accuracy. However, even advanced imaging may fail to

visualize the appendix in cases of agenesis or atypical anatomical positions, leading to diagnostic confusion and unnecessary surgical intervention^{9,11}.

Case reports from diverse geographical regions underscore this diagnostic challenge. Published cases include young adults and adolescents in whom classic symptoms prompted surgical exploration, only to reveal absence of the appendix intraoperatively. For example, appendiceal agenesis was diagnosed in a 25 year old pregnant woman explored for suspected appendicitis, and in a 19 year old female investigated for inflammatory appendiceal syndrome, with both cases revealing no appendicular tissue on exploration^{8,18}. Another case series from Africa reported agenesis in 0.001% of laparotomies for presumed appendicitis, while isolated reports describe the absence of the appendix in routine cadaveric dissection, suggesting its occurrence even in asymptomatic individuals^{13,14,18}.

Intraoperative recognition of appendiceal agenesis requires a methodical and careful approach. When the appendix cannot be located in its expected position, surgeons should trace the taeniae coli meticulously from the caecum, mobilize the caecum, and dissect the visceral peritoneum carefully to exclude retrocecal, pelvic, or subhepatic positions. Failure to conduct a comprehensive search may lead to misdiagnosis, inadvertent injury to the caecum or surrounding bowel, or inappropriate surgical escalation. A reported complication includes iatrogenic caecal perforation during extensive dissection to locate an absent appendix, necessitating right hemicolectomy¹⁹.

The presence of concurrent pathologies, especially in female patients, further complicates diagnosis. Gynecological conditions such as ovarian torsion, ruptured ovarian cysts, pelvic inflammatory disease, and endometriosis can closely mimic appendicitis and may only be identified intraoperatively or on postoperative imaging. In such contexts, failure to identify the appendix coupled with discovery of alternative pathology underscores the need for broad differential diagnosis and careful intraoperative evaluation^{9,16}.

Historically, both open and laparoscopic approaches have been employed for evaluation of suspected appendicitis. Laparoscopy, in particular, offers diagnostic advantage by permitting direct visualization of the entire abdominal cavity and adnexal structures, potentially reducing unnecessary laparotomies and postoperative morbidity. However, limitations include availability, expertise, and visualization challenges in patients with extensive adhesions or obesity. Traditional open exploration remains relevant in many settings, especially where laparoscopy is not feasible.

Without proper diagnosis and appropriate treatment, clinicians risk surgical complications, delayed management of the true underlying condition, and increased morbidity. Misdiagnosis can lead to unnecessary appendectomies, missed treatment of the actual pathology, prolonged hospital stays, and increased healthcare costs. Therefore, maintaining awareness of rare anatomical anomalies is essential for optimal surgical decision-making.

Future advances in diagnostic technology hold promise for improving preoperative detection of anatomical anomalies such as agenesis. Artificial intelligence-assisted interpretation of CT and MRI imaging could enhance sensitivity for atypical or absent appendicular structures. Genetic and molecular research into developmental signaling pathways may also elucidate the embryological basis of rare congenital anomalies, potentially allowing for earlier and more precise diagnosis.

In the present case, despite classical symptoms of appendicitis and preliminary imaging suggestive of appendiceal inflammation, intraoperative exploration failed to reveal the appendix. A systematic search of all anatomical regions did not identify the organ, corroborated by postoperative imaging, and alternative pathology was pursued and treated appropriately. This experience underscores the importance of comprehensive assessment, interdisciplinary understanding of embryological and Ayurvedic principles, and careful operative strategy when encountering rare presentations such as appendiceal agenesis.

Overall, while appendiceal agenesis is exceptionally rare, its recognition is clinically important to avoid unnecessary procedures and to identify alternate causes of acute abdomen, particularly in female patients. Multimodal evaluation and continued reporting of such cases will enhance understanding and surgical outcomes.

Conclusions

Agenesis of the veriform appendix is an exceptionally rare anomaly that can closely mimic the clinical presentation of acute appendicitis, creating significant diagnostic and surgical challenges. Surgeons may encounter classical signs and radiological findings suggestive of appendicitis, yet intraoperative exploration may reveal complete absence of the appendix, highlighting the importance of careful inspection of the cecal region. Awareness of this rare variation is crucial for avoiding misdiagnosis and unnecessary interventions. Recognizing such anomalies, along with meticulous operative assessment, helps ensure accurate management and optimal patient outcomes, even in the face of unexpected anatomical variations.

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Declaration of patient consent

Authors certify that they have obtained a patient consent form, where the patient has given his consent for reporting the case along with the images and other clinical information in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflicts of Interest- No any.

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