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Clinical Case Study

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Schwannoma of Common Bile Duct: A Clinico-Radiologic Diagnostic Quagmire – A Case Report

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Keywords
Schwannoma · Benign nerve sheath tumor · Common bile duct · Porta hepatis

Abstract
Background: Schwannomas are benign nerve sheath tumors that are extremely rare in the biliary tract. A comprehensive review of literature enumerated approximately 30 case reports of schwannoma in the biliary tract tree and porta hepatis region. ***Case Presentation:*** We report a case of a 40-year-old female who presented with abdominal pain. Imaging revealed a mass at the porta hepatis extending from the portal bifurcation till the hilum encasing the main portal vein and abutting the right portal vein. Differentials of carcinoma, lymphoma, and mesenchymal tumor were kept. Ultrasound-guided biopsy of the mass showed a benign nerve sheath tumor, immunopositive for S100. The histopathological evaluation of the excised mass confirmed the origin of mass in the common bile duct. ***Conclusions:*** Our case highlights that schwannomas, though benign, can mimic a carcinoma or lymphoma if present at a rare site such as bile ducts. An exhaustive clinical and radiological workup with diligent histopathological evaluation is mandatory in dealing with such rare cases as radical surgery and chemotherapy can be avoided in such patients.

Schwanoma da via biliar biliar comum: Um diagnóstico clinico-patologico volúvel

Palavras Chave
Schwannoma · Tumor benigno da bainha nervosa · CBD · Porta hepatis

Resumo
Introdução: Os schwannomas são tumores benignos da bainhas nervosas, que são extremamente raros ao nível das vias biliares. Uma revisão abrangente da literatura enumerou cerca de 30 casos de schwannomas com envolvimento da árvore biliar e da região da Porta Hepatis. ***Apresentação do caso:*** Relatamos um caso de uma doente de 40 anos que apresentava dor abdominal. A imagem revelou uma massa que se prolonga desde a bifurcação da veia porta até ao hilo hepático, com

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"encasement" da veia porta principal e "abutement" da veia porta direita. Foram considerados os diagnósticos diferenciais de carcinoma, linfoma e tumor mesenquimatoso. A biópsia guiada por ecografia da massa mostrou um tumor benigno da bainha nervosa, imunopositivo para o S100. A avaliação histopatológica da massa excisada confirmou a sua origem na via biliar comum. ***Conclusões:*** O nosso caso realça que os schwannomas, embora benignos, podem imitar um carcinoma ou linfoma se estiverem presentes num local raro, como os canais biliares. Um trabalho clínico e radiológico exaustivo com uma avaliação histopatológica diligente é obrigatória para orientar com casos tão raros, em que a cirurgia radical e a quimioterapia podem ser evitadas.

Background

Schwannomas are benign encapsulated nerve sheath tumors, usually attached to peripheral nerves and arise from differentiated Schwann cells. Mostly, they are sporadic, while some are associated with syndromes such as neurofibromatosis type 2, schwannomatosis, or Carney's complex [1]. These are spindle cell tumors that generally occur in the upper limbs, head, and neck, followed by the trunk and flexor surfaces of the lower extremities. Approximately 0.2% of all gastrointestinal (GI) tumors are constituted by schwannomas [2]. The commonest site is the stomach followed by the colon, cecum, and rectum and rarely the jejunum [3]. GI schwannomas are uncommon and usually occur in sixth to seventh decade [4]. Herein, we report a rare case of schwannoma of the common bile duct (CBD) presenting as a porta hepatis mass. Few case reports of porta hepatis schwannomas arising from CBD, hepatoduodenal ligament, hepatic vein, or artery have been published in the literature.

Case Presentation

A 40-year-old female, presented with the chief complaint of pain in the abdomen for the past 10 months. Pain was located in the right upper quadrant, dull in nature with no aggravating factors. There was no history of fever, jaundice, vomiting, and upper or lower GI bleeding. The patient did not report any altered bowel habits, loss of appetite, or weight. Patient had no medical comorbidities. The general physical and abdominal examination was unremarkable. Complete hemogram and renal function tests were within normal limits. SGOT and SGPT were 26 and 25 IU/L, respectively. Serum bilirubin was 1.2 mg% while alkaline phosphatase was 186 IU/L.

Ultrasonography (USG) of the abdomen showed a hypoechoic mass at the porta abutting the right portal vein and the main portal vein (shown in Fig. 1). The liver was normal with no intrahepatic biliary radicle dilation and showed a normal echotexture. No ascites was reported. Contrast-enhanced computed tomography abdomen revealed a mass at the porta extending from the portal bifurcation till the hilum encasing the main portal vein and abutting the right portal vein (shown in Fig. 1). Common hepatic artery was free. Contrast-enhanced magnetic resonance imaging (MRI) abdomen showed a mass at the porta abutting the right portal vein, main portal vein, right hepatic artery, and the common hepatic artery (shown in Fig. 1). The mass was extending between the head of the pancreas and the inferior vena cava. Vertically, the mass was extending between the hilum and retropancreatic region. Endoscopic ultrasound was also done which revealed a mass from the superior mesenteric vein/portal vein confluence till the hilum (shown in Fig. 2). Fat planes with the main portal vein were maintained. Based on overall clinical and imaging findings, possibilities of carcinoma, lymphoma, and mesenchymal tumor were considered.

USG-guided biopsy was performed on the mass which showed a benign nerve sheath tumor, immunopositive for S100 (shown in Fig. 3). Following the above investigations, the patient was undertaken for surgery. En bloc excision of the mass and CBD with Roux-en-Y hepatico-jejunostomy was performed. The resection specimen was submitted to the department of pathology. Gross examination showed an encapsulated, circumscribed, yellow-white firm tumor measuring 7.5 cm in maximum dimension (shown in Fig. 3). On microscopy, it was a biphasic tumor composed of hypercellular areas with fascicular arrangement of spindle cells and palisades (Verocay bodies) along with myxoid hypocellular areas and focal hyalinization. The spindle-shaped tumor cells contained moderately ill-defined cytoplasm, wavy tapering nuclei with fine granular nuclear chromatin, and inconspicuous nucleoli. In addition, there were many interspersed blood vessels with hyalinized walls along with lymphoid infiltrate at the tumor periphery (shown in Fig. 3). No significant nuclear pleomorphism, mitosis, or necrosis was noted. On immunohistochemistry, the tumor cells were immunopositive for vimentin, S100, and SOX10 while they were negative for CK, CD34, smooth muscle actin, CD117, DOG1, myogenin, ALK, STAT6, CD21, and HMB45. Ki67 proliferation index was less than 1% (shown in Fig. 4). Based on the immunohistomorphological profile, the tumor was diagnosed as schwannoma. Patient recovered well postoperatively and was discharged on postoperative day 10. The patient is disease-free on follow-up after 36 months.

Discussion

Schwannomas in porta hepatis and biliary tree are very rare and approximately 30 cases have been reported in literature till date. There are 22 reported cases of schwannoma in the biliary tract alone details of which have been summarized in Table 1 [3–24].

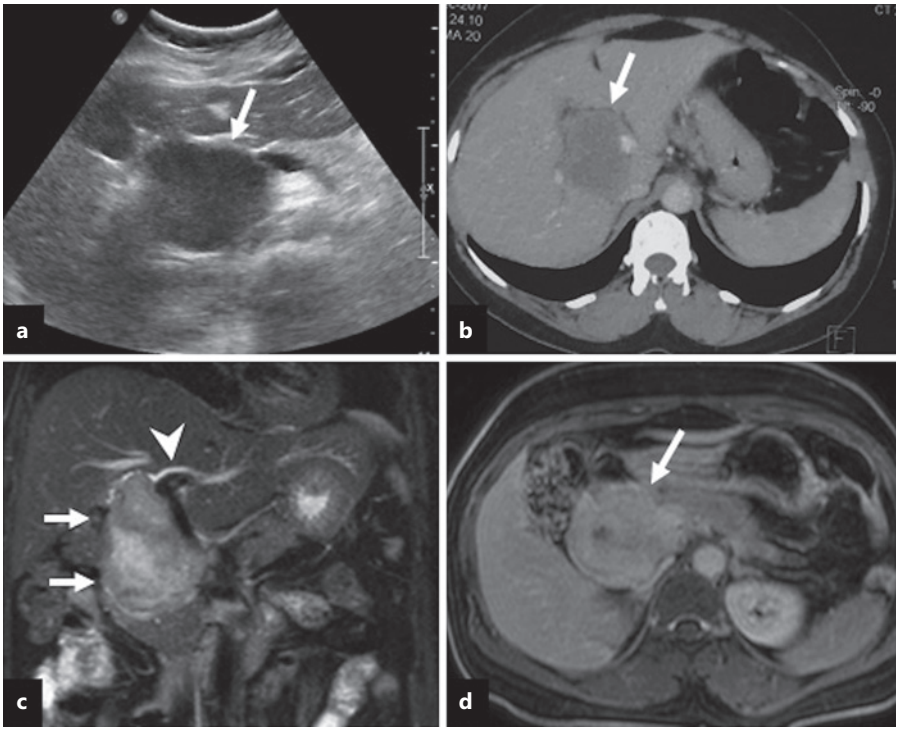

Fig. 1. a Ultrasound image shows a hypoechoic mass (arrow) at the porta hepatis. **b** Axial contrast-enhanced CT image shows a hypodense mass (arrow) at the porta hepatic splaying the portal veins. **c** Coronal T2-weighted MR image shows an oblong hyperintense mass (arrows) along the course of the CBD with mild intrahepatic bile duct dilatation (arrowhead). **d** Axial contrast-enhanced T1-weighted MR image (5 min delayed) shows late enhancement of the mass (arrow).

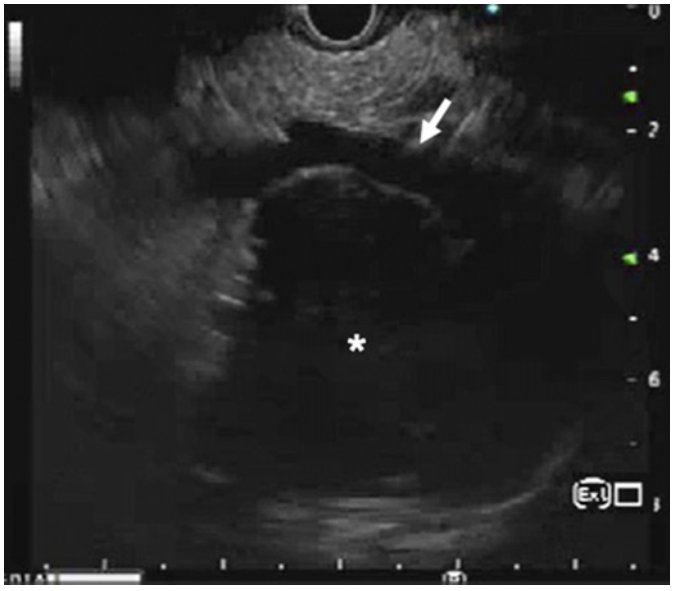

Fig. 2. Endoscopic ultrasound shows a hypoechoic mass (asterisk) at the porta hepatis abutting the portal vein (arrow). Image courtesy of: Dr Deepak Gunjan.

A significant female predominance with male-to-female ratio of 1:5.5 was seen in these patients. The mean age at presentation was 48.3 years (range 15–78 years). Abdominal pain and jaundice were the most common presenting symptoms in these patients (jaundice in 38%, abdominal pain in 19%, both pain and jaundice in 23% of patients, respectively). The most common location was CBD (57%). The preoperative clinico-radiological diagnoses were quite variable and comprised metastatic melanoma, gastrointestinal stromal tumor (GIST), lymphoma, adenocarcinoma, leiomyosarcoma.

In all the cases, it was extremely difficult to correctly diagnose this tumor preoperatively, mainly due to the fact that tumors at this site can easily mimic bile duct adenocarcinoma and other malignancies such as lymphoma and IgG4-related diseases. Moreover, the location is difficult to approach for a minimally invasive technique such as fine needle aspirate. Surgical resection was carried out in most cases with unremarkable postoperative period [3–24]. In our case, a correct preoperative diagnosis on USG-guided biopsy helped in the adequate surgical management.

Radiologically, contrast-enhanced MRI is better suited to visualize the extent and size of such soft tissue tumors and dilatation of bile duct radicles due to mass effect [25]. On MRI, schwannomas are hypointense on T1-weighted images and homogeneously hyperintense on T2-weighted images [26]. Usually, degenerative

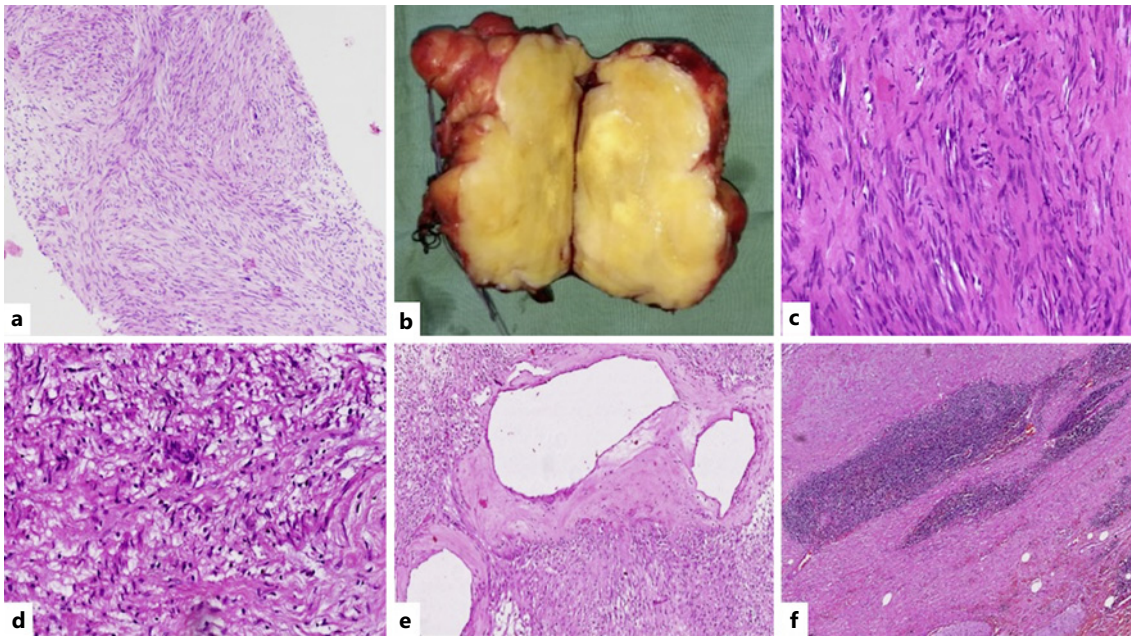

Fig. 3. a Biopsy section shows a benign spindle cell tumor (×100, H&E). **b** Gross examination shows nodular circumscribed yellow-white firm tumor. The tumor shows hypercellular areas with Verocay bodies (**c**) (×400, H&E) and hypocellular areas (**d**) (×400, H&E). Also, many dilated vessels with perivascular hyalinization (**e**) (×200, H&E) and lymphoid cuff at tumor periphery (**f**) (×200, H&E) are seen.

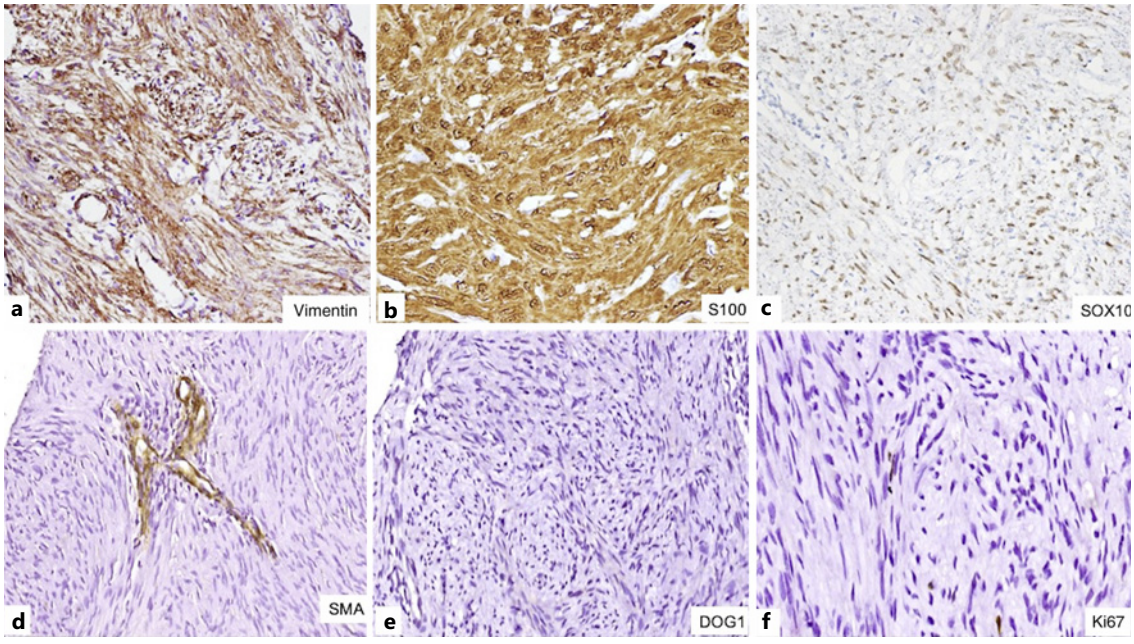

Fig. 4. The tumor cells are immunopositive for vimentin (**a**), S100 (**b**), and SOX10 (**c**) while they are immunonegative for SMA (**d**), DOG1 (**e**), and show low Ki67 proliferation index (**f**) (×200). SMA, smooth muscle actin.

Table 1. Details of previously reported cases of schwannoma in the biliary tract [3–24]

No.	Authors	Age, years/sex	Site	Presentation	Treatment	Outcome
1	Oden et al. 1955 [3]	40/F	CBD	Jaundice	SR	DF
2	Whisnant et al. 1974 [4]	15/F	Distal CBD	Abdominal pain, jaundice	SR	DF (12 months)
3	Balart et al. 1983 [5]	56/F	EHBD	Pain, jaundice	SR	DF
4	Honjo et al. 2003 [6]	48/F	CBD	Jaundice	EL + SR	DF (36 months)
5	Jakobs et al. 2003 [7]	37/M	CBD	Jaundice	SR	DF (12 months)
6	Otani et al. 2005 [8]	59/F	Remnant choledochal cyst	Abdominal pain	SR	DF (15 years)
7	Vyas et al. 2006 [9]	29/F	CBD	Jaundice	SR	DF (12 months)
8	Park et al. 2006 [10]	53/F	Porta hepatis (CBD)	Asymptomatic	EL + SR	DF (11 months)
9	Kamani et al. 2007 [11]	39/F	EHBD	Jaundice	SR	DF
10	Fenoglio et al. 2007 [12]	41/F	CBD	Pruritus, weight loss	EL + SR	DF (12 months)
11	Jung et al. 2007 [13]	64/F	EHBD	Asymptomatic	SR	DF
12	Madhusudan et al. 2009 [14]	46/F	IHBD & EHBD	Jaundice	No SR	-
13	Kulkarni et al. 2009 [15]	38/F	CBD	Abdominal pain and jaundice	SR	DF (3 months)
14	De Sena et al. 2009 [16]	58/F	EHBD	Jaundice	NA	NA
15	Parameshwarappa et al. 2010 [17]	38/F	CBD	Abdominal pain, jaundice	EL + SR	DF (12 months)
16	Panait et al. 2011 [18]	54/F	CHD	GERD	EL + SR	DF
17	Fonseca et al. 2012 [19]	64/F	EHBD	Incidental	Localized SR	DF (12 months)
18	Campos et al. 2016 [20]	62/M	IHBD	Abdominal pain, jaundice	CHDR	DF (18 months)
19	Xu et al. 2016 [21]	31/F	IHBD and EHBD, GB	Abdominal pain, abdominal distension	SR	DF (70 months)
20	Kolhe et al. 2019 [22]	46/F	CBD	Jaundice	NA	NA
21	Takami et al. 2021 [23]	78/M	EHBD	Incidental finding	EHBDR	DF
22	Ishimaru et al. 2021 [24]	68/M	Lower CBD	Abdominal pain	Local BDR with cholecystectomy	DF (30 months)
23	Present case	40/F	CBD	Abdominal pain	SR	DF (36 months)

CHDR, common hepatic duct resection; DF, disease free; EHBD, extrahepatic bile duct; EHBDR, extrahepatic bile duct resection; EL, exploratory laparotomy; SR, surgical resection.

Table 2. Discussion of common differential diagnoses of soft tissue masses in the biliary tract region

Differential diagnosis	Histomorphological features	Benign/ malignant	Immunohistochemistry
Neurofibroma	Less cellular, spindle cells with no significant nuclear pleomorphism, no mitosis or necrosis	Benign	S100+ (strong), SOX10+ (strong), CD34+ (fingerprint-like positivity)
Granular cell tumor	Cells with abundant granular cytoplasm in sheets	Benign	S100+
Leiomyoma	Spindle cells with blunt nuclear ends and cigar-shaped nuclei in fascicles, no significant nuclear pleomorphism, mitosis, or necrosis	Benign	SMA+, desmin+, h-caldesmon+, SMMHC+
GIST	Cellular tumors, mostly spindle cells in fascicles and sheets, sometimes epithelioid cells, mild to moderate nuclear pleomorphism, variable mitosis and necrosis	Benign/ malignant	CD117+, DOG1+, CD34+/−, SMA+/−, S100+/−
Leiomyosarcoma	Cellular tumors composed of spindle cells with blunt nuclear ends and cigar-shaped nuclei in fascicles, significant nuclear pleomorphism, mitoses, and necrosis	Malignant	SMA+, desmin+, h-caldesmon+, SMMHC+, high Ki67

SMA, smooth muscle actin; SMMHC, smooth muscle myosin heavy chain.

changes are uncommon in GI schwannomas, but if present can lead to error in diagnosis. Diagnosis in such cases can only be made on histology which requires excision of the mass. The most common differential diagnoses for schwannoma are other benign and malignant soft tissue tumors of the gastrointestinal tract such as neurofibroma, leiomyoma, GIST, and leiomyosarcoma, enumerated in Table 2.

Histologically, schwannomas show hypercellular areas (Antoni A) with Verocay bodies and hypocellular (Antoni B) areas. The tumor cells are predominantly spindle-shaped with hyperchromatic nuclei. Mitosis and necrosis are usually absent [10]. Schwannomas can show an array of degenerative changes such as hyalinization, calcification, hemorrhage, myxoid change, cyst formation, focal bizarre nuclear atypia. Multiple variants are seen, namely, ancient, plexiform, cellular, epithelioid, microcystic or reticular, and melanotic or pigmented [14].

Immunohistochemistry plays an important role in distinguishing schwannoma from close differentials such as GIST, leiomyomas, or neurofibromas. Schwannomas are strongly immunopositive for S100 protein and SOX10. GISTs are immunopositive for c-KIT and DOG1, while leiomyoma and leiomyosarcoma will show strong and diffuse smooth muscle actin positivity. CD34 and calretinin might help in differentiating these from neurofibroma as both are immunopositive for S100. Neurofibroma shows moderate CD34 positivity. Fine et al. [27] in their study compared calretinin positivity in schwannoma and neurofibroma and 96% of schwannomas displayed strong calretinin staining compared to only 7% in neurofibroma.

GI schwannomas show some histomorphological variations when compared to schwannomas at other regions in the form of predominance of hypercellular areas (Antoni A) and lack of nuclear palisading pattern as seen in conventional schwannoma [28]. Our case, although it showed both the areas, Lasota et al. [29] also reported lack of *NF-2* gene alterations in the GI schwannomas suggesting it to be a morphologically and genetically distinct group of nerve sheath tumors. However, more data needs to be incorporated to arrive successfully at this conclusion.

Being benign tumors, these have an excellent prognosis and complete surgical resection is the mainstay of treatment. Recurrence or malignant transformation is rarely seen [30].

Conclusion

CBD schwannomas are very rare and a surgeon's nightmare until the histopathological examination is done. The present case adds to the rare list of schwannomas arising from CBD and also enumerates the diagnostic challenges from a pathologist's perspective.

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Statement of Ethics

Study approval from the institute's Ethics Committee (AIIMS, New Delhi) was not required. A written informed consent was obtained from the participant for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Shilpi Thakur contributed to manuscript writing, editing, and data curation. Adarsh Barwad and Prasenjit Das contributed to review and editing. Nihar Ranjan Dash and Kumble S. Madhusudhan helped with clinical and radiological data curation. Rajni Yadav reviewed and edited the manuscript.

Data Availability Statement

No data were generated during this study. Further inquiries can be directed to the corresponding author.

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