
CASE REPORT**Mesenteric Microcystic/Reticular Schwannoma: A Diagnostic Dilemma***Sandhya S. Ware^{1*}, Dilip S. Sarate¹, Ajay D. Jungare¹, Vinod K. Sawaitul¹**¹Department of Pathology, Government Medical College, Akola-444001
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Abstract:

Microcystic/reticular schwannoma is a rare variant of schwannoma with a predilection for visceral locations including the gastrointestinal tract. Due to overlapping features with other tumours, unawareness of this tumour type may lead to diagnostic pitfalls. We here report a case of microcystic/reticular schwannoma arising in the mesentery of a 25-year-old female. The tumour was incidentally discovered by ultrasonography. An operation was performed and histological examination revealed a capsulated tumour composed of spindle cells with reticular pattern set within a myxoid stroma. Differential diagnoses were Extra Gastrointestinal Stromal Tumour (EGIST) with myxoid change, microcystic/reticular schwannoma, Low-Grade Fibromyxoid Sarcoma (LGFMS) and Aggressive Angiomyxoma (AAM). Immunohistochemically, the tumour cells were positive for S-100 protein which was consistent with a peripheral nerve sheath tumour. Albeit very rare, microcystic/reticular schwannoma should be included in the differential diagnosis of mesenteric tumours. The awareness and the knowledge about this rare tumour are needed to achieve the correct diagnosis.

Keywords: Aggressive Angiomyxoma, Extra-Gastrointestinal Stromal Tumour, Low-Grade Fibromyxoid Sarcoma, Mesenteric Tumours, Microcystic/Reticular Schwannoma

Introduction:

Schwannoma is a benign neurogenic tumour. It occurs in the head and neck, extremities, mediastinum, retroperitoneum, and pelvis. Mesenteric schwannoma is extremely rare where it is difficult to diagnose [1, 2]. The typical schwannomas are characterized by alternating Antony A and Antony B areas. In addition to the classic type, approximately 11 morphological variants have been recognized [3]. Microcystic/reticular schwannoma represents a distinctive variant of schwannoma, initially described in 2008 and shows a predilection for visceral location including gastrointestinal tract [1, 4].

We present here a rare case of microcystic/reticular schwannoma arising in the mesentery in a young female.

Case Report:

A 25-year-old female presented with intermittent abdominal pain since six months. Past and family history was insignificant. On general examination patient had mild anemia. Local examination revealed tenderness in right lower abdomen. A mass was palpated at the middle abdomen. Routine laboratory investigations were within normal limits except for anemia. Patient underwent Ultrasonography (USG), followed by

CT scan. USG revealed mass with solid and cystic areas approximately 18×13 cm in lower abdomen. USG could not detect exact origin of the mass. CT scan images revealed a large, well circumscribed, heterogeneously enhancing, soft tissue mass in the pelvis, superior to the uterus showing few calcified foci within and measuring approximately 13×9×13 cm in size. Based on the clinical and radiological findings, differential diagnosis of Extra-Gastrointestinal Stromal Tumour (EGIST) and pedunculated subserosal uterine fibroid were provided.

Patient underwent laparotomy. The mass was removed “en-bloc” with piece of small intestine attached to it. It weighed 600 gm and the mass was sent for histopathological examination. The submitted specimen comprised of 14cm long piece of jejunum with a capsulated mass; 13cms in diameter attached to the mesentery (Fig. 1). Cut sections showed whitish yellow, homogenously solid, gelatinous mass with a myxoid appearance (Fig.2). Few areas of chalky white calcifications were seen. Histological examination revealed a tumour surrounded by fibrous capsule with a reticular architecture. It was composed of relatively alternating fibrillary and myxoid areas (Fig.3).The tumour cells were slender spindle shaped with eosinophilic cytoplasm arranged in anastomosing and intersecting strands in a lace like reticular and microcystic pattern. The nuclei were oval and tapered and showed inconspicuous nucleoli (Fig. 4). There was no palisading or Verocay body formation. Features suggestive of malignancy were absent.

Immunohistochemically, the tumour cells were positive for S-100 protein (Fig. 5) and negative for CD117, Desmin, and smooth muscle actin, CD34,

ER and PR. Thus, on histopathology and immunohistochemistry, a diagnosis of mesenteric microcystic/reticular schwannoma was given.



Fig. 1: Specimen Composed of Jejunum with a Capsulated Mass Arising from Mesentery



Fig. 2: Gross Cut Section showing Capsulated Tumour with Whitish Yellow, Gelatinous Myxoid Mass

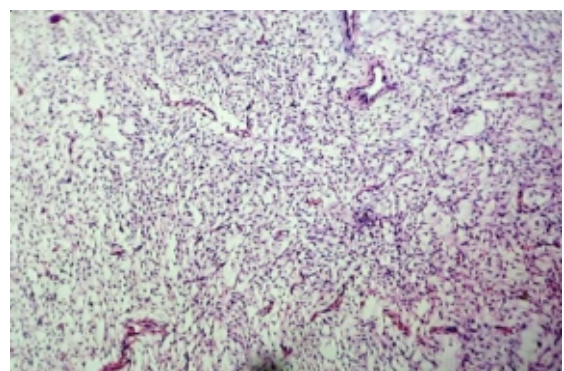


Fig. 3: H& E Stained Section showing Tumour Composed of Fibrillary and Myxoid Areas

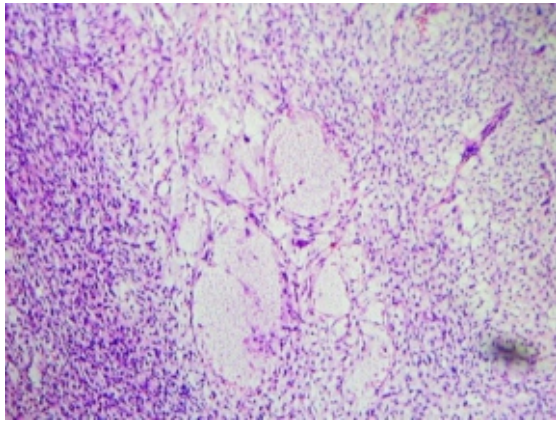


Fig. 4: H & E Stained Section showing Bipolar Spindle Shaped Cells with Microcystic Areas

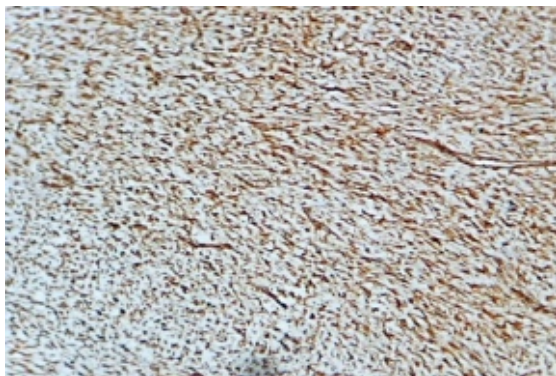


Fig. 5: Tumour Cells showing Diffuse Nuclear and Cytoplasmic Positivity of S-100 Protein
Discussion

Schwannoma is an encapsulated, slow growing neoplasm which can be intracranial, in the spine, or extracranial. Mesenteric schwannoma is rare. Microcystic/reticular schwannoma is a rare variant of schwannoma [2]. Due to its rarity and overlapping features with other wide variety of tumours, unawareness of this tumour type may lead to diagnostic and therapeutic pitfalls. Therefore, to enhance its awareness, we present here a case of mesenteric microcystic/reticular schwannoma.

On histology, microcystic/reticular schwannoma differs from a classic schwannoma in many ways. The former lacks the Antony A and Antony B

areas, presence of palisading or Verocay bodies, aggregates of foamy histiocytes, and hyalinized blood vessels. The hallmark of microcystic/reticular schwannoma is the presence of a striking reticular and microcystic architecture, a feature not observed in any other variant of schwannoma. Regardless of the preferential location in the gastrointestinal tract, microcystic/reticular schwannoma is different from so-called gastrointestinal schwannoma. Gastrointestinal schwannoma is relatively more common than microcystic/reticular schwannoma. It is more common in stomach. Histologically, it is composed of spindle cells in microtrabecular pattern with peritumoural lymphocytic cuff [3].

Besides variant of schwannoma, there are other spindle cell tumours with myxoid change occurring in mesentery which may cause confusion. These tumours include EGIST with myxoid change, Aggressive Angiomyxoma (AAM) and Low-Grade Fibromyxoid Sarcoma (LGFMS) [3].

EGIST are rare mesenchymal tumours that originate outside the gastrointestinal tract, commonly mesentery, omentum and retroperitoneum. They are more common in patients over the age of 50 years. Grossly, it varies from firm, fleshy gray large masses to cystic ones. Myxoid change is seen in majority, associated with extensive hemorrhage or necrosis [5]. Histopathologically, majority of EGIST are composed of spindle cells and few of epithelioid cells. Microcystic change can be seen in EGIST mimicking microcystic/reticular schwannoma. Immunohistochemically, EGIST are positive for CD117 and DOG1, whereas negative for S-100 protein. As the treatment varies greatly, a distinction between EGIST and microcystic/reticular schwannoma is warranted [3].

AAM is a rare soft tissue tumour originating from myofibroblasts. It is a benign but locally infiltrative tumour. It usually arises in the pelvic and perineal organs in women of reproductive age group. Extra-genital AAM have been reported in the literature. This tumour can be rarely encapsulated. Grossly, it is bulky with a gelatinous cut surface. Microscopically, it can mimic microcystic/reticular schwannoma. Immunohistochemically, AAMs show positivity for vimentin, desmin, smooth muscle actin, ER and PR [6].

LGFMS is a rare mesenchymal tumour. It arises from soft tissues of the lower extremities. Abdominal LGFMS is extremely rare. Microscopically, it is composed of spindle cells in myxoid and fibrotic stroma, but has a high rate of recurrence and metastasis. LGFMS is negative for S-100[7]. Diagnosis of microcystic/reticular schwannoma should always be considered in

cases where myxoid sarcomas are suspected, thereby avoiding aggressive intervention and overtreatment.

A histopathology with immunohistochemistry, help in distinguishing these lesions from other similar entities. In our case, the tumour cells were positive for S-100. Thus, the tumour was of peripheral nerve sheath origin and histologically, microcystic/reticular schwannoma.

Mesenteric microcystic/reticular schwannoma may cause a diagnostic dilemma, especially in a third world setting where diagnosis is hampered by lack of facilities and poverty. Albeit very rare, it is important to entertain microcystic/reticular schwannoma in the differential diagnosis of mesenteric tumours. We report this case to highlight its existence and enhance pathologists and clinicians awareness to avoid misdiagnosis and mistreatment.

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