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Mesenteric Microcystic/Reticular Schwannoma: A Diagnostic Dilemma

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ABSTRACT

Background:- Microcystic/ reticular schwannomais a rare variant of schwannoma with a predilection for visceral locations including the gastrointestinal tract. Due to overlapping features with other tumors, unawareness of this tumor type may lead to diagnostic pitfalls. Case:- We here report a case of microcystic/reticular schwannoma arising in the mesentery of a 25-year-old female. The tumor was incidentally discovered by ultrasonography. Observations & Result:- An operation was performed and histological examination revealed a capsulated tumor composed of spindle cells with reticular pattern set within a myxoid stroma. Differential diagnoses were Extra gastrointestinal stromal tumor with myxoid change (EGIST), Microcystic/ reticular schwannoma, low-grade fibromyxoid sarcoma (LGFMS) and aggressive angiomyxoma (AAM). Immunohistochemically, the tumor cells were positive for S-100 protein which was consistent with a peripheral nerve sheath tumor.

Conclusion:- Albeit very rare, Microcystic/ reticular schwannoma should be included in the differential diagnosis of mesenteric tumors. The awareness and knowledge about this tumor are needed to achieve the correct diagnosis.

KEYWORDS : Microcystic/reticular schwannoma, Extra-gastrointestinal stromal tumor, Low-grade fibromyxoid sarcoma, Mesentric tumors

INTRODUCTION:-

Schwannoma is a benign, generally nonrecurring neurogenic tumor. It occurs predominantly in the soft tissues of the head and neck, extremities, mediastinum, retroperitoneum, and pelvis with no sex predilection. Mesenteric schwannoma is extremely rare where it is difficult to diagnose[1,2].

The typical schwannomas are characterized by a biphasic pattern composed of alternating Antony A and Antony B areas. In addition to the classic type, approximately 11 morphological variants have been recognized. These variants include ancient [degenerated] schwannoma, cellular schwannoma, plexiform schwannoma, melantoicschwannoma, epitheliodschwannoma, hybrid schwannoma/neurofibroma, hybrid schwannoma/perineuroma, gastrointestinal schwannoma, neuroblastoma like schwannoma represents a distinctive morphologic 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 DETAILS:-

A 25 year old female presented with complaints of intermittent diffuse abdominal pain since 5 to 6 months. There was no history of fever, constipation or malaena. Past and family history was insignificant. On general examination patient was vitally stable with mild anemia. Local examination revealed mild tenderness in right lower abdomen. The abdomen was distended, and a mass was palpated at the middle abdomen. Other systemic examination revealed no significant abnormality.

Routine laboratory investigations were within normal limits except for anemia. Patient underwent ultrasonography (USG), which was then followed up by with CT scan. Ultrasonographic (USG) examination revealed mass lesion with solid and cystic areas approximately 18x13 cm in lower abdomen. USG could not detect exact origin of mass. CT scan images revealed a large, well defined, well circumscribed, heterogeneously enhancing, necrotic soft tissue mass in the pelvis, superior to the uterus extending cranially in the abdominal cavity up to the umbilical region, showing few calcified foci within and measuring approximately 13x9x13 cm in its maximum dimensions. The radiologic features were suggestive of neoplastic etiology, possibly arising from bowel loops. Based on the clinical and radiological findings, differential diagnosis of Extra-gastrointestinal stromal tumor (EGIST) and pedunculated subserosal uterine fibroid were provided.

OBSERVATION & RESULT:-

Patient underwent laparotomy. Intraoperative findings revealed solid, well circumscribed, encapsulated mass measuring approximately 13 cm in diameter within the mesentery of the jejunum. The mass was removed "en-bloc" with piece of small intestine attached to it; end to end anastomosis was done. It weighed 600gm and the mass was sent for histopathological examination. The patient recovered uneventfully after surgery.

The submitted specimen comprised of 14 cm long piece of jejunum with a well circumscribed, capsulated mass; 13 cms in diameter attached to the mesentery. Cut sections showed whitish yellow, homogenously solid, gelatinous, and soft torubbery mass with a myxoid appearance showing no features of hemorrhage or necrosis. Few areas of chalky white calcification were seen. Histological examination revealed a well circumscribed tumor surrounded by a thin fibrous capsule with a reticular architecture. It was composed of relatively alternating fibrillary and myxoidareas. The tumor cells were slender bipolar spindle shaped with eosinophillic cytoplasm arranged in anastomosing and intersecting strands in a lace like reticular and microcystic pattern set within a myxoid stroma. The nuclei were round, oval and tapered and showed inconspicuous nucleoli. There was no palisading or Verocay body formation. Features suggestive of malignancy, like nuclear pleomorphism, mitosis or necrosis were absent.

Immunohistochemically, the tumor cells showed diffuse and strong nuclear and cytoplasmic staining of S-100 protein which was consistent with a peripheral nerve sheath tumor. Tumor cells were negative for CD117, Desmin, smooth muscle actin, CD34, ER and PR. Thus, on histopathology and immunohistochemistry, a diagnosis of mesenteric microcystic/reticular schwannoma was given.

DISCUSSION:-

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

Taking a review of literature for mesenteric microcystic/reticular schwannoma, there was a predilection for female patients. Clinically the majority of patients presented with an asymptomatic mass detected incidentally by imaging examination and few patients complained of change in bowel habit, pain[5,6,7].

On histology, microcystic/reticular schwannoma differs from schwannoma in many ways. The former lacks the distinctive features of a classic schwannoma. Namely alternating areas of Antony A and Antony B, 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 microcysticarchitecture, a feature not observed in any other variant of schwannoma[3]. Regardless of preferential location in the gastrointestinal tract, microcystic/reticular schwannoma is different from other variant of so-called gastrointestinal schwannoma. Gastrointestinal schwannoma is relatively more common than microcystic/reticular schwannoma. This type of schwannoma tends to occur in stomach, although intestine and mesentery can be occasionally involved. Histologically, it is composed of spindled Schwann cells displaying frequently a microtrabecular pattern and characterized by peritumoral lymphocytic cuff. Although focal myxoid change can occur in a few gastrointestinal schwannoma, prominent microcystic pattern is never seen[3].

Besides variant of schwannoma, there are other types of spindle cell tumors with myxoid change occurring in mesentery which may cause confusion among them. These tumors include Extra-gastrointestinal stromal tumor (EGIST) with myxoid change, low-grade fibromyxoid sarcoma (LGFMS) and aggressive angiomyxoma (AAM)[3].

Extra-gastrointestinal stromal tumor (EGIST) are rare mesenchymal tumors that originate outside the gastrointestinal tract, commonly mesentery, omentum and retroperitoneum. Furthermore, EGIST tend to be more common in patients over the age of 50 years. Gross appearance of EGIST varies from firm, fleshy gray-red large masses to cystic once. Cystic and myxoid change is seen in majority, assocated with extensive hemorrhage or necrosis[8]. Histopathologically, majority of EGIST are composed of spindle cells and few of epithelioid cells. Cystoplasmic vacuolization, mucin deposition and microcystic change can be seen in EGIST mimicking microcystic/reticular schwannoma. Immunohistochemically. EGIST are positive for CD117 and DOGI1, whereas the staining of S-100 protein is consistently negative. It is worthy to note here that focal immunoractivity of CD117 can be observed in minority of microcystic/reticular schwannoma. As the treatment varies greatly, a distinction between EGIST and microcystic/reticular schwannoma is warranted[3].

Aggressive angiomyxoma (AAM) is a rare myxoid soft tissue tumor originating from fibroblasts/myofibroblasts. It is a distinct, slow growing, benign but locally infiltrative tumor. It usually arises in the pelvic and perineal organs, mostly in women in reproductive age group. Extra-genital aggressive angiomyxoma involving the axilla, the chest wall, oral cavity and supraclavicular fossa have been reprted in the literature. This tumor can be rarely encapsulated suggesting an isolated mesenchymal cell origin. Grossly, it is bulky with a glistening, gelatinous cut surface. Microscopically, it can mimic microcystic/reticular schwannoma. Immunohistochemically, AAMs show positivity for vimentin, desmin and smooth muscle actin. The immunoractivity of the estrogen (ER) and progesterone (PR) receptors have been described to be the most characteristic feature[9].

Low-grade fibromyxoid sarcoma (LGFMS) is a rare, deceptively benign mesenchymal tumor. It usually arises from deep soft tissues of the lower extremities, but occasionally reported to arise also, from mediastinum, inguinal region, neck, buttock and brain. Abdominal LGFMS is extremely rare, it had been reported to arise from small bowel mesentery, colon, retroperitoneum and the ovary. Microscopically, this tumor is composed of bland spindled to stellate cells in myxoid and fibrotic stroma, but has fully aggressive behavior and high rate of recurrence and metastasis. LGFMS is negative for S-100[10]. Diagnosis of microcystic/ reticular schwannoma should always be considered in cases where myxoid sarcomas are suspected as it can mimic myxoid sarcomas, clinically and radiologically, thereby avoiding aggressive intervention and overtreatment.

CONCLUSION:-

A histopathology combined with immunohistochemistry, help in distinguishing these lesions from the more commonly occurring similar entities. In our case, the tumor cells were diffusely positive for S-100 and negative for CD117, desmin, smooth muscle actin, CD34, ER, PR. Thus, the tumor was of peripheral nerve sheath origin histologically, microcystic/ reticular schwannoma.

Mesenteric microcystic/ reticular schwannoma may cause a diagnostic dilemma, especially in a third world setting where preoperative diagnosis is hampered by lack of facilities and poverty. A high index of suspicion is needed for preoperative diagnosis for proper planning of the operation. Albeit very rare, it is important to entertain microcystic/ reticular schwannoma in the differential diagnosis of mesenteric tumors. Awareness of the entity and an immunohistochemical panel of markers will ensure that the correct diagnosis is made. We report this case to highlight its existence and enhance pathologist's and clinicians awareness of this under-recognised variant of schwannoma so as to avoid misdiagnosis and mistreatment.

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