

Cellular Schwannoma Arising from Sigmoid Mesocolon Presenting as Torsion

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Abstract

Schwannomas are a type of peripheral nerve sheath tumors with clinically indolent behavior. Though, they can occur anywhere in body, the incidence in retroperitoneum, mediastinum, and pelvis is exceedingly rare. We present a case of a 58-year-old female with a massive twisted tumor arising from sigmoid mesocolon. The tumor was diagnosed to be a case of cellular schwannoma, an exceedingly rare tumor in this location with rare presentation.

Keywords: Pelvis, Retroperitoneum, Schwannoma, Sigmoid mesocolon, Torsion, Tumor

Introduction

Schwannomas are derived from neural crest derived Schwann cells.^[1] Most common sites of occurrence of schwannomas are head, neck, and extremities. Most of these tumors are painless, despite their neuronal origin. Usually there is no associated symptom, unless and until the tumor grows large, and causes compression of adjacent organs. The retroperitoneum can harbor a wide range of lesions, including rare benign and malignant neoplasms that can be either primary or metastatic lesions.^[2] Schwannomas account for 1-10% of all primary retroperitoneal tumors and up to 0.7% of benign schwannomas and 1.7% of malignant schwannomas arise in retroperitoneum.^[3]

Case Report

A 58-year-old female presented to emergency department with acute abdomen of 2 h onset. Clinically, the patient was afebrile and had tenderness in lower abdomen. Ultrasonography (USG) and emergency plain computer tomography (CT) scan was carried out. USG revealed a homogenous and hypoechoic solid lesion in pelvis without any evidence of lymphadenopathy. Plain CT displayed a large pelvic mass of approximate size of 17 cm × 15 cm × 12 cm extending to lower abdomen, with

mass effect causing displacement of the bowel superiorly and toward right. Mass appeared relatively dense in plain CT. Right ovary could not be visualized and left ovary appeared bulky. There was moderate degree of ascites and minimal pleural effusion. Based on clinical symptoms and plain CT findings, a presumptive diagnosis of torsion of right ovarian neoplasm was made. Emergency laparotomy was carried out. During laparotomy, it was evident that the large mass was arising from sigmoid mesocolon and was twisted around its axis with marked congestion on its surface. Complete surgical excision was carried out. Histopathological examination of the specimen was carried out. Cut surface [Figure 1] of the specimen revealed well encapsulated lesion containing multiple nodules of grey white solid areas and areas of congestion. Microscopic examination [Figure 2] of multiple sections from grey white solid areas revealed fascicles, whorls and palisades (Antoni A areas) of spindle cells varying from wavy nerve bundles to plump elongated cells with moderate eosinophilic cytoplasm. Verocay bodies were absent and Antoni B areas were scanty. Dense hyalinization was evident in all the sections studied. Small to large caliber hyalinized congested blood vessels were seen. Immunostaining showed diffuse S-100 positivity [Figure 2, inset] and no reactivity for CD117, CD34, SMA, CD31, HMB-45, and desmin. A diagnosis of cellular schwannoma with torsion was conferred.

Discussion

The schwannomas are typically found as solitary tumors. Presentation associated with schwannomatosis or neurofibromatosis type 2 (NF2) has been noted. Most schwannomas are benign, clinically silent, but rarely malignant changes can occur, in association with NF2.^[4] Grossly schwannomas are well encapsulated solid tumors with

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Figure 1: Cut surface of a well encapsulated tumor shows grey white nodules, areas of congestion and cystic changes

smooth surface and are ovoid or spherical in shape. Rarely secondary changes such as cystic degeneration, hemorrhage, and calcification can occur.^[4] Pre-operative diagnosis of schwannomas can be difficult. Various radiological examinations such as USG, CT scan or angiography fail to differentiate schwannomas from other retroperitoneal tumor. Diagnosis is based on histopathological examination and immunohistochemistry. Cystic degeneration and torsion of tumor as seen in present case evokes sharp pain. Microscopically dimorphic pattern is usually the characteristic feature of schwannomas with cellular Antoni A region and loose myxoid Antoni B regions. Cellular schwannomas are uncommon variants of schwannomas (approximately 5% of benign peripheral nerve sheath tumors) that are often misdiagnosed for malignancy because of deep location, hypercellularity and mild degree of nuclear pleomorphism.^[5] The common location of cellular schwannomas are soft tissues such as retroperitoneum, posterior mediastinum, pelvis and also bone.^[6] Cellular schwannomas are type of schwannomas with predominant Antoni A region and lack of verocay bodies, increased nuclear pleomorphism and low mitotic figures (up to 4/10 high power field). Antoni B area may be present, but not usually exceeding 10% of the lesion, as seen in the present case. Cellular schwannomas are also characterized by presence of hyalinized thick vessels, clusters of foamy histiocytes, thick capsule, and lymphoid aggregates may be seen at places. They may be mistaken for malignant nerve sheath tumors or malignant spindle cell tumor such as leiomyosarcoma, fibrosarcoma, gastrointestinal stromal tumor, metastatic melanoma, Kaposi's sarcoma, solitary fibrous tumor, inflammatory fibroid polyps, and inflammatory myofibroblastic tumors.^[5] For differentiation of cellular schwannomas from these tumors, immunohistochemical staining for various markers such as CD117, CD34, SMA, desmin, S-100, CD31, and HMB-45 is necessary. These schwannomas exhibit strong diffuse S-100 immunoreactivity, unlike that of malignant peripheral nerve sheath tumor and negative staining for all

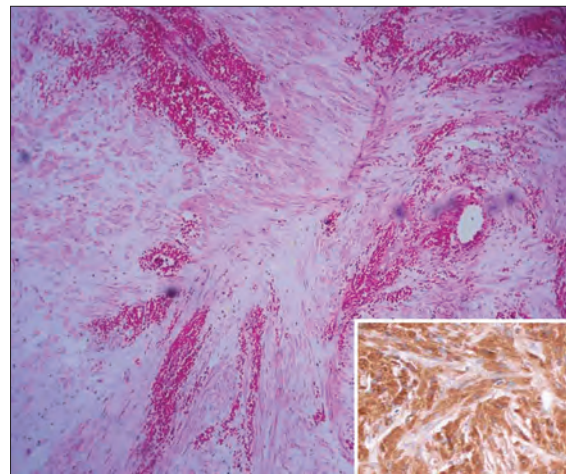


Figure 2: Section shows cellular Antoni A areas and areas of hemorrhage (H and E stained section, $\times 100$). Inset shows diffuse S-100 immunoreactivity

the above listed markers. Schwannomas can rarely undergo torsion and infarction^[7] and present with acute lower abdomen, as seen in the present case. Total excision of schwannoma is therapeutic and has a good prognosis. Incomplete excision can lead to recurrence of tumor.

The present case reflected the variable presentation of rare type of schwannoma in a rare location, misdiagnosed clinically and radiologically as torsion of ovarian neoplasm.

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How to cite this article: ????

Source of Support: Nil. Conflict of Interest: None declared.