Primary Benign Schwannoma of the pleura: A rare case

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ABSTRACT

Schwannoma, also called neurilemmoma is a benign peripheral nerve sheath tumor. Most common locations are flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots and cerebellopontine angle. Pleural schwannomas are extremely rare neoplasms of the thoracic cavity. To the best of our knowledge, till date, less than 20 cases have been reported. Herein, we report a case of primary benign pleural schwannoma in an elderly 60 year old female in whom the pleural mass was discovered incidentally on CT chest. Patient was subjected to right postero-lateral thoracotomy, pleural mass was resected which showed histopathological and immunohistochemical features of schwannoma.

KEYWORDS: Pleura, thoracic cavity, schwannoma.

INTRODUCTION

Schwannoma is an encapsulated benign peripheral nerve sheath tumor which is almost always solitary. Common locations are the flexor surfaces of the extremities, neck, mediastinum, retroperitoneum, posterior spinal roots and cerebellopontine angle.[1] Pleural schwannomas are rare and arise from the autonomic nerve fibre sheaths in the pleura. They are generally benign, asymptomatic, slow growing lesions and occur more commonly in adults with a male predominance.

Histopathological differential diagnosis includes neurofibroma and other benign spindle cell tumors like leiomyoma, leiomyosarcoma and calcifying aponeurotic fibroma which may also show palisading of the nuclei.[2] Histopathological variants of this lesion are ancient schwannoma, benign myxoid schwannoma, benign epithelioid schwannoma, benign glandular schwannoma and psammomatosusmelanotic schwannoma.[2]

CASE REPORT

A 60 year old elderly female presented with respiratory symptoms due to acute exacerbation of asthma. Patient is a known seasonal asthmatic since 10 years. Physical examination revealed bilateral wheeze. All routine laboratory investigations were within normal limits. CT scan chest showed evidence of well defined, cystic mass with partial peripheral enhancement measuring 6x4cm involving the right side of posterior mediastinum. No pleural effusion/thickening observed(Figure 1).

A radiological diagnosis of “posterior mediastinal cystic mass- Neurofibroma” was considered. Patient was posted for surgery and a right postero-lateral thoracotomy was done. A mass was seen arising posteriorly from the pleura, measuring 7 cm x 5 cm, which was partly solid and cystic. Excision of pleural mass was done and specimen sent for histopathological examination.

Gross examination of the specimen revealed grey white solid and membranous soft tissue bits partly cystic, showing mucoid and haemorrhagic areas.

Microscopic sections studied from the specimen revealed a well encapsulated mass showing benign spindle cells with alternating hypercellular Antoni A areas with nuclear palisading with vascularity, bodies formation and hypocellular Antoni B areas showing myxoid changes and thick walled blood vessels with perivascular hyalination. No mitosis or necrosis was noted. A provisional diagnosis of schwannoma was considered.

IHC stain S-100 performed showed diffuse and strong positivity of tumor cells. Based on histopathology and IHC,a final diagnosis of primary pleural benign peripheral
A nerve sheath tumor consistent with schwannoma was made. Patient had an uneventful postoperative recovery. Postoperatively, patient received no adjuvant therapy. 6 months later, on follow up, patient was completely asymptomatic and with no evidence of tumor recurrence.

Figure 1: CT scan showing SOL in the posterior mediastinum

Figure 2: H&E 10x view showing schwannoma with hypercellular areas and verocay bodies

Figure 3: IHC S-100-10x view showing strong positivity

DISCUSSION

Primary pleural schwannomas are exceedingly rare neoplasms [3]. They account for 1-2% of all thoracic tumors[4]. Although it can occur at any age, pleural schwannomas commonly affect adults between their third and sixth decades. In addition, males are more commonly affected than females [5].

Majority of pleural schwannoma are benign, though malignancy has been reported in some cases [6]. Schwannomas arise from specialized myelin producing cells-Schwann cells, of either the sympathetic or parasympathetic autonomic nerve fibre sheaths [7]. Schwannomas arising in the pleural surface of the lung generally grow slowly, hence they do not usually produce symptoms. The majority of patients with pleural schwannomas are often asymptomatic or may present with non-specific and vague symptoms. Therefore, the vast majority of pleural schwannomas are discovered incidentally during investigations for other complaints. Although they are rarely encountered, malignant pleural schwannomas have been reported in the literature [8]. Malignant lesions have been described in patients with neurofibromatosis type 1 and patients with positive history of previous radiation therapy [9]. In addition, malignant pleural schwannomas are likely to produce symptoms due to their large size compared to benign lesions. Large tumors have the potential to produce pain and neurological symptoms.
symptoms due to their mass occupying effect and compression on adjacent structures [10].

Diagnosing pleural schwannomas is often challenging. Definitive diagnosis cannot be reached even with advanced imaging modalities and laboratory investigations. Radiological images aid in raising the suspicion about the nature of the lesion and narrowing the differential diagnosis. Pleural schwannomas should be included in the differential diagnosis of solitary, solid, well demarcated pleural lesions, which include, but are not restricted to, pleural lipomas, pleural metastasis, mesotheliomas, and solitary fibrous tumors. Moreover, laboratory tests usually lie within normal limits. Therefore, definitive diagnosis can only be established through histopathological examination and immunohistochemical staining of the neoplasm, which requires a section of the tumor.

CT scan remains the diagnostic imaging modality of choice for these neoplasms. CT scan can outline the size, number and exact location of the lesions. CT scan can also demonstrate cystic and/or solid components of the tumor. Malignant pleural schwannomas have similar features on CT scan, however they are usually associated with presence of pleural nodules, pleural effusions and metastatic pulmonary nodules [11].

Microscopically, Antoni A and Antoni B areas are revealed in the majority of pleural schwannoma cases. Antoni A represents areas of hypercellularity with verocay bodies. Antoni B areas of myxoid hypocellularity exhibit degenerating changes (i.e. cyst formation, haemorrhage, calcification, xanthomatous infiltration and hyalinisation) [12]. Immunohistochemically, pleural schwannomas typically stain diffusely and strongly positive for S-100 protein.

The standard care of management of pleural schwannomas is primarily surgical resection thoracoscopically or complete pleural resection of the tumor whenever technically possible with frequent continuous follow up.

REFERENCES


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