Posterior Mediastinal Castleman's Disease: A Case Report

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Abstract Castleman's disease is a rare lymphoproliferative disorder, which usually occurs in the chest. The castleman's disease is often asymptomatic, but it can cause nonspecific thoracic symptoms such as cough and dyspnea. Surgical removal is curative and malignant transformation has not been described. We report an unusual case of Castleman's disease who presented as a posterior mediastinal mass with review of the relevant literature.

Keywords Castleman's disease · Posterior mediastinum

Abbreviations

CT Computed tomography
T6 Thoracic level 6

Introduction

Castleman's disease, also known as "angiofollicular lymph node hyperplasia," "giant lymph node hyperplasia," and "angiomatous lymphoid hamartoma," is an uncommon lymphoproliferative disorder, which most frequently occurs as an asymptomatic mass in the mediastinum [1]. The

disease was first described by Castleman in 1954 as a form of "benign lymph node hyperplasia simulating thymoma" [2]. 70% of cases of Castleman's disease are located in the chest, along the tracheobronchial tree in the mediastinum or lung hilus, although it can occur in other areas such as the pelvis, neck, retroperitoneum, and muscle [3]. We report an unusual case of Castleman's disease which presented as a posterior mediastinal mass.

Case Report

A 30 years female patient presented to our institute with complaints of Lt sided chestpain and backache for 3 months. Her medical history and medical examination were normal. Her blood counts and full biochemical profile was also normal. Her Chest X-ray and CT thorax (Fig. 1) revealed a well defined lobulated soft tissue density lesion of $9.6 \times 7 \times 6.5$ cm in the posterior mediastinum in left paravertebral location extending from T6-T9 vertebral levels. The lesion showed moderate homogenous enhancement with no rib erosion/vertebral erosion/paratracheal/hilar lymphadenopathy. There was displacement of left lower lobe bronchus with adjacent segmental atelectasis.

We performed a left lateral thoracotomy and on exploration an encapsulated $10 \times 7 \times 7$ cm mass was present in the costovertebral sulcus covered by parietal pleura. The tumour was located in the paravertebral area in the posterior mediastinum and was firmly adherent to the chest wall. The entire tumour was removed along with its capsule and soft tissue. The lesion was hypervascular deriving its blood supply from the intercostal vessels. Histopatholog-

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Fig. 1 Computed tomography scan of the chest showing a well-defined densely homogeneous tumor arising in the sixth left intercostal space

ically, the tumor composed of lymphoid follicles which contained multiple capillaries surrounded by hyaline sheaths (Fig. 2). The microscopic features were characteristic of the hyaline vascular type seen in Castleman's disease. The patient was well when last seen 3 months after her operation, asymptomatic. Chest X-ray showed no evidence of recurrence.

Discussion

Since Castleman et al. first reported a case of "localised mediastinal lymph node hyperplasia resembling thymoma," similar cases have been reported as "Castleman's disease" [2, 4]. Although most of these lesions occur within the chest, other regions, including the neck, pelvis, retroperitoneum, and axilla, may be involved [3, 4]. There is no remarkable sex predominance or identifiable risk factors in

the development of Castleman's disease, [3, 4] which has been reported to arise as a tumor inflammation or hamartoma, but its cause is still unknown [5, 6]. Three histological types have been described, namely, hyaline vascular, plasma cell, and mixed type [3, 7]. Patients are usually either asymptomatic or have nonspecific symptoms such as cough, dyspnea, chest pain, respiratory infection, and back pain, [3, 4] mainly caused by tracheobronchial compression.

The diagnosis of Castleman's disease is ultimately made by histology, requiring either removal or biopsy of the lesion for definitive diagnosis. The clinical manifestation of the disease is highly dependent on the histopathology. The plasma cell type can bring about more severe symptoms than the hyaline-vascular type and has a strong association with extrathoracic, multicentric, and highly aggressive disease [8]. Chest wall involvement by Castleman's disease is a very rare manifestation. Rena et al. found only five reports in an 11 year review of the literature of Castleman's disease, adding their patient as the sixth case [3]. Thereafter; Erdogan et al. reported the seventh case. Therefore, to the best of our knowledge our patient represents only the eighth case [9].

According to our review of these eight patients, two were men and six were women; six had nonspecific symptoms and two were asymptomatic; and thoracotomy was performed for seven patients and sternotomy for one. Histological picture showed mixed type in only one (0.125%) patient, and the hyaline vascular type in the other seven (87.5%). Symptomatic relief was achieved by surgery in all of the patients. There was no report of recurrence or malignancy after excision of Castleman's disease involving the chest wall [3]. Thoracotomy is necessary for Castleman's disease involving the chest wall because needle biopsy has low diagnostic accuracy and thoracoscopic biopsy is too dangerous for this highly vascular tumor [9].

Fig. 2 Histopathologically, the tumor was composed of lymphoid follicles that contained multiple capillaries surrounded by hyaline sheaths **a** low power, **b** high power

Conclusion

Castleman's disease presenting as a posterior mediastinal mass is a rare entity. Thoracotomy and excision of the mass is the treatment of choice, both as a diagnostic as well as therapeutic modality since the diagnosis of Castleman's disease is made only on histology. Recurrence after excision of the mass is not known.

Consent Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests The authors declare that they have no competing interests.

Author's contributions SS-1 wrote the original manuscript and interpreted the patient data with regard to the haematological and radiological diagnosis. SS-2 analysed the data. All the authors have read and approved the final manuscript.

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