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Cervico-Thoracic Approach for a Large Desmoid Tumor

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Abstract

Intrathoracic desmoid tumors are very rare. We report a case of a 35-year-old woman, who had dyspnea and dysphagia. The imaging studies showed a voluminous mass tissue occupying the thoracic outlet and extending to the medium mediastinum. A scanno-guided percutaneous needle biopsy initially concluded to a solitary fibrous tumor of the pleura. Surgery was decided. The approach of this voluminous apical and intra thoracic mass was challenging. Complete resection was successfully performed by Cormier Dartevelle Grunenwald's route. Final pathology was consistent with desmoid fibromatosis. The post-operative course was uneventful.

Keywords: Pleura • Surgery • Immunohistochemistry • Cormier Dartevelle Grunenwald • Desmoid fibromatosis

Introduction

Desmoid fibromatosis are very rare mesenchymal tumors with a partially aggressive growth pattern and high relapse rates, without metastases. Intrathoracic desmoid tumors are unusual. Among therapeutic alternatives, wide resection is the mainstream treatment [1]. Although, the adequate approach could be a real challenge for voluminous cervico-thoracic tumors. We report the case of a large desmoid tumor with unusual localization.

Case Report

We report a case of a 35-year-old woman, with no past medical history complaining from dyspnea and dysphagia since 3 months. There was no history of cough or weight loss. Chest x-ray revealed an apical opacity occupying two-thirds of the left hemithorax (Figure 1a). A contrast enhanced computed tomography scan of the thorax showed a voluminous well encapsulated mass on the thoracic outlet extending to the medium mediastinum repressing the left lung.

The subclavian vessels seemed to be invaded. (Figures 1b and 1c). MR Angiography revealed a heterogeneous well limited mass compressing the subclavian vein without invasion of the subclavian artery neither the brachial plexus. (Figure 1d). Preliminary pathology from scannoguided biopsy and immunohistochemistry (PS 100 negative) showed a benign tumor consistent with a solitary fibrous tumor. Wide surgical resection was decided in a multidisciplinary consultation meeting.

The question was how to approach this voluminous apical pleural mass and to have both cervical and thoracic access. As the mass has a similar presentation of apical tumors, the chosen approach was Cormier Dartevelle Grunenwald route. (Figures 2a and 3a).

The tumor fills the left apex and plunges into the thorax, it encompasses the artery and the subclavian vein and it invaded the 1st rib. The 1st rib was cut. The left subclavian artery was progressively dissected free. The subclavian vein was invaded, it was controlled and sectioned. The apical part of the lesion was released progressively and the whole tumor was mobilized. After assessing the tumor extirpability, a complementary thoracotomy did not seem

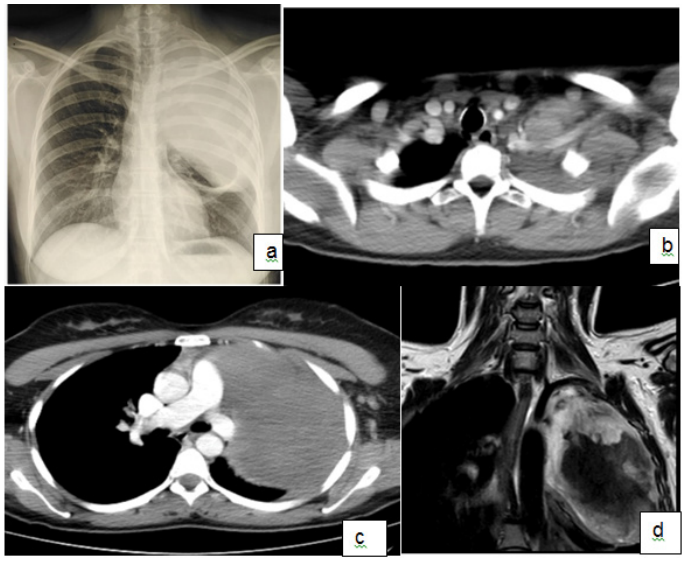

Figure 1. (a) Chest X ray demonstrated a large apical opacity extending to the left chest. (b and c) Chest CT scan showed the mass in the region of the left apex surrounding the subclavian vessels and protruding on the left pleural cavity compressing the left lung. (d) At MRI the subclavian artery was compressed by the mass and the brachial plexus was not infiltrated.

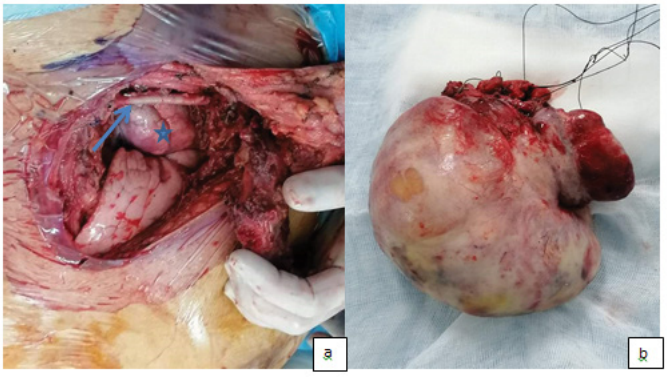

Figure 2. (a) Per-operative photos; the star shows the tumor, the rest of the mass is hidden by the lung, The arrow showing the left subclavian artery well dissected. (b)The surgical specimen.

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to be necessary. The voluminous mass was then carefully delivered through the cervical incision (Figures 2a and 2b).

The postoperative course was complicated by a left chylothorax treated initially by fat free diet and long-acting somatostatin analog and then re-operated. She had exclusive videothoracoscopy for thoracic duct ligation with uneventful follow-up.

Final pathology of the surgical specimen concluded to desmoid type fibromatosis. Microscopically, the mass consisted of elongated slender spindle cells with uniform appearance surrounded by abundant collagen without atypical nuclei neither mitosis with a low proliferation index Ki 67. The cells were immuno-stained with monoclonal antibodies for ß-nuclear catenin but not for S100 protein, CD34, SOX 10, MUC 4 and STAT 6. These results have ruled out the diagnosis of solitary fibrous tumor and malignant peripheral nerve sheaths tumor. At the last follow up, at one year, the CT scan showed no tumor recurrence.

Discussion

Desmoid fibromatosis are rare tumors accounting approximately 0.03% of all neoplasms. They are characterized by proliferation of fibroblast and myofibroblast-type spindle cells infiltrating musculo aponevrotic tissues [2]. They occur sporadically or in the course of familial adenomatous polyposis. Those tumors are reported to affect different ages between 15 and 60 years with female predilection [2]. Thoracic localization presents about 20% of all extra abdominal desmoids. They generally affect the chest wall most commonly in the shoulder girdle [2].

Intra thoracic desmoid tumor, like our case, is an extremely rare entity described in the literature as a tumor that originates from the pleura or mediastinum in the thoracic skeleton with minimal chest wall involvement [3]. True intra thoracic desmoids remains asymptomatic until the tumor becomes too large, with the surrounding organs [3]. In our case the main symptom was dyspnea and the tumor sizes 11 cm. It was diagnosed preoperatively as a solitary fibrous tumor of the pleura since its fibroblastic origin and its benign histologic appearance.

This case is very interesting in that it underlines intra thoracic desmoid tumor as a differential diagnosis of spindle cell proliferations of the pleura including low grade sarcoma and solitary fibrous tumor. Immunohistochemistry can help differentiate between these tumors [4].

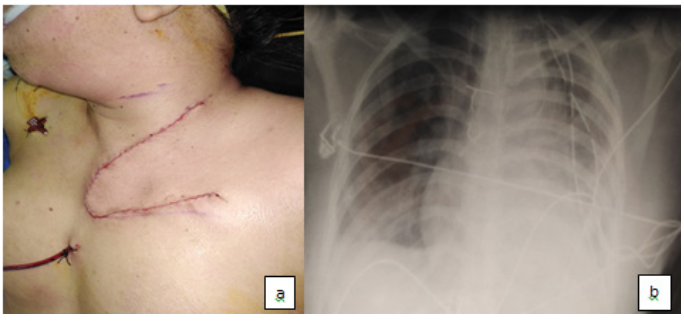

Figure 3. (a) Post-operative scar. (b) Post-operative chest X-ray showing a good re-expansion of the lung.

Intra thoracic desmoid tumors are considered borderline with aggressive local behavior. Their management is challenging. Wide surgical resection is the treatment of choice of resectable tumors. Surgery should be discussed in multidisciplinary consultation meeting referring to radiological outcomes [5].

CT scan and MRI allow defining mass limits and neighbor's organs eventual. MRI accurately evaluates the relationship between soft tissue tumors and other structures, such as bone, vessels, and nerves. Typical desmoid-type fibromatosis shows isosignal on T1WI, high signal on T2WI, and strong heterogeneous enhancement on enhanced T1WI [6].

One bloc resection via the adequate route is difficult for the surgeon especially for large Cervico-thoracic tumors. In our case the Cormier Dartevelle Grunewald approach offered an excellent access to the sub-clavian region with a safe control of neurovascular outlet structures. Fortunately, the tumor was successfully removed from the cervical approach without using anterior thoracotomy. Complete resection with free margins is an essential prognostic factor and recurrences are strongly associated with macroscopically positive margins [2]. The efficacy of radiotherapy when surgical margins are negative is controversial and not proved. Adjuvant radiotherapy should be considered for incompletely resected tumor or local recurrence [1,2,5]. A long term follow up is necessary to check on time eventual recurrence [1].

Conclusion

Intra thoracic desmoid tumors are exceptional. A multidisciplinary management is necessary to treat this rare entity. Wide resection via the suitable approach is challenging and correlated with prompt and distant outcomes.

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