A rare chest wall tumor: A case report of angiomatosis

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Abstract
Angiomatosis, a pathology that may occur in any soft tissue, is typically characterized by vascular proliferation and invasion of adjacent tissues. Although it is described as a benign lesion, it can be as challenging as malignancy in treatment and follow-up due to its vascular rich structure and frequent recurrence. Here, we present a rare case of chest wall angiomatosis in a 46-year-old female patient treated with surgical excision.

Keywords: Angiomatosis, Mediastinum, Thoracic Wall.

INTRODUCTION

Angiomatosis, a benign vascular pathology commonly seen in the first two decades of life, can involve any soft tissue, such as breast, bone, and retina (1). It is a very rare type of tumor and, although benign, recurrence rate after surgery is reported at approximately 90%. The standard surgical approach is resection with wide margins, which requires diligent preoperative planning (2). Here, we report an angiomatosis case treated with complete chest wall resection.

CASE REPORT

A 46-year-old female patient with abdominal pain for one month had a chest X-ray in another health facility (Figure 1A). He was referred to our clinic after abdominal computed tomography (CT) scan revealed vascular branches extending from the aorta to the tumor suggesting sequestration (Figure 1B). Moreover, chest-wall soft-tissue density in the left paravertebral region in the cross-sectional area was observed on thorax CT (Figure 1C). On the CT scan, vascular branches extending from the aorta to the tumor were observed, suggesting sequestration (Figure 1C). A positron emission tomography (PET) scan showed a 70X27 mm tumor on the left lateral of the T9-T11 vertebrae, extending from the 10th and 11th intercostal spaces to the paraspinal musculature with a maximum standard uptake value of 4.02. Transthoracic fine-needle aspiration biopsy yielded a fibroadipose tissue sample containing very few spindle cells, which was non-diagnostic. Due to its rich vascular structure, it was consulted for preoperative embolization, but the procedure was not considered appropriate by interventional radiology. Then after forced expiratory volume in 1 second value (1.93, 89%) in pulmonary function test was deemed suitable for surgical resection, an exploratory left thoracotomy uncovered a tumor, extending from the paravertebral region to the ribs, that was highly hemorrhagic even in frozen section biopsy. However, the frozen section examination did not establish whether the tumor was benign or malignant. Therefore, the decision was made to proceed with chest wall resection. We observed many collateral branches from the aorta to the tumor and ligated these during the excision. The 9th, 10th, and 11th ribs were disarticulated posteriorly and they were resected anteriorly at a distance of 4 cm from the tumor, followed by en bloc resection of the tumor (Figure 2). Afterwards chest wall reconstruction using a Gore-Tex patch was performed since the soft tissue is also resected together with the ribs, in order to avoid chest wall instability. On pathological examination, the tumor was invading the surrounding fibroadipose tissue without evident mitosis or necrosis was positive for CD31 and CD34. No invasion of the ribs was detected. In light of these findings, we confirmed the diagnosis of angiomatosis (Figure 3). The patient, who had no postoperative chemotherapy (CT) and radiotherapy (RT), is currently in the 3rd year of follow-up without recurrence.

Figure 1. A. Preoperative chest X-ray, B. Abdominal CT Scan (Vascular structures originating from the aorta), C. Abdominal CT Scan (Soft tissue invasion), D. Postoperative chest X-ray

Figure 2. Macroscopic view of the tumor
DISCUSSION

Angiomatosis is a benign tumor histologically characterized by vascular proliferation and invasion of adjacent fibroadipose tissue. The first reported case of angiomatosis in 1927 was retinal, and the disease was later shown to involve almost any soft tissue. These scarce types of tumors most commonly occur in the lower extremity, followed by the chest wall, abdomen, and upper extremity (4).

The main limitation in the preoperative diagnosis of angiomatosis lies in the fact that the pathology includes extensive fibroadipose tissue and dilated vascular structures, as well as necrosis. Typically, percutaneous biopsy does not yield a precise diagnosis, as seen in our case and others in the literature (1,5). When the studies in the literature were examined, it was seen that the biopsy result was not diagnostic and was not associated with malignancy in two separate thoracic wall case reports (3,4). Due to the presence of abnormal vascular structures and the likelihood of hemorrhage in angiomatosis, an imaging-guided biopsy is particularly recommended for large tumors that appear irresectable. The definitive diagnosis is based on evident vascular proliferation in gross pathology specimens (5). In our case, neither the frozen section examination nor preoperative interventions proved diagnostic.

Although angiomatosis is defined as a vascular proliferation with asymptomatic bone destruction, we observed no bone tissue invasion in our case despite the presence of bone tissues within the tumor region. The standard treatment in angiomatosis is wide surgical resection. Due to fibroadipose tissue invasion, which was also present in our case, high recurrence rates are reported for the tumor, necessitating wide surgical margins (6). Besides, preoperative embolization can reduce the tumor size before surgery since the tumor is vascular and may involve considerable proliferation. We also believe that preoperative tumor embolization could contribute to facilitate the operation in our case since we observed many collateral branches originating from the aorta, and the tumor was hemorrhagic. However, radiotherapy and chemotherapy are among the treatment approaches in cases unsuitable for surgery (7).

To conclude, angiomatosis is a scarce type of tumor that cannot be diagnosed preoperatively and, albeit benign, is frequently recurrent and invasive. Therefore, surgical en bloc resection and close follow-up can be recommended as the standard treatment approach.

Declarations

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Ethical Committee approval was not required. Informed consent was obtained from all participants.

REFERENCES
