Case Report

An unusual cause of bleeding: angiomatoid fibrous histiocytoma: a case report

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ABSTRACT

Angiomatoid fibrous histiocytoma (AFH) is a rare mesenchymal neoplasm that typically affects the skin and subcutaneous tissues. It is often painless and with slow growth. We present a case of a 57-year-old female patient with an unusual lesion diagnosed as AFH, located in the abdominal wall, manifested clinically with bleeding. The patient underwent exploratory laparotomy with an excision of the mass (apparently preperitoneal) and hepatic subsegmentectomy II, for histological exam. This case is noteworthy due to its uncommon location and clinical presentation.

Keywords: Angiomatoid fibrous histiocytoma, Mesenchymal neoplasm, Bleeding, Hepatic subsegmentectomy

INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is an uncommon tumor found in soft tissues, displaying intermediate biological behavior and uncertain differentiation. It predominantly manifests in the extremities of children and young adults. In 2013, the World Health Organization (WHO) initially categorized Angiomatoid fibrous histiocytoma (AFH) as an intermediate tumor with uncertain differentiation.

Although it displays specific histological characteristics, including a nodular arrangement of oval and spindle cells, cystic cavities containing blood, and a dense surrounding lymphoplasmocytic infiltrate, the morphological spectrum of Angiomatoid fibrous histiocytoma (AFH) is extensive. The uncommon nature of its occurrence and the lack of a distinct immunohistochemical profile can pose challenges in making an accurate diagnosis.

Currently, AFH is recognized in an increasing number of anatomical sites. The behavior of AFH is indolent and surgery is the main treatment and is generally curative.

CASE REPORT

A 57-year-old woman with a history of breast cancer who had undergone mastectomy four years earlier, no usual medication, presented to the Emergency Department with upper abdominal pain, asthenia, and cutaneous pallor. There was no history of abdominal trauma or recent abdominal surgeries.

Physical examination revealed a mass and tenderness upon palpation of the upper abdominal wall, without peritoneal irritation.

Laboratory tests showed a hemoglobin level of 8 g/dl.
Figure 1: Mass of the upper abdominal wall.

A computed tomography revealed a large hematoma on the anterior abdominal wall in an apparent epigastric pre-peritoneal position, measuring about 99×68 mm, with evidence of active bleeding originated from a branch of the right inferior epigastric artery. She was hemodynamically stable.

Figure 2: Computed tomography showing large hematoma on the anterior abdominal.

The case was discussed with Interventional Radiology, which performed arterial embolization.

Further evaluation excluded hematologic causes as the etiology of this apparently spontaneous hematoma.

During hospitalization, the patient remained stable with a rise in hemoglobin following embolization, leading to her discharge at day 13 for continued outpatient evaluation.

A repeat abdominal CT scan two months later showed a hematoma with overlapping characteristics and size compared to the previous one. A biopsy was performed, for a suspected hypervascularized mass in its capsule, which returned negative for malignant cells. Subsequently, (after 3 months of maintaining the mass/hematoma with de same size) a multidisciplinary team decided on surgical intervention.

The patient underwent exploratory laparotomy with an excision of the mass (apparently preperitoneal) and hepatic subsegmentectomy II, for histological exam due to alterations underlying the mass, in that region.

Figure 3: Mesenchimal lesion.

Figure 4: After resection.

The patient was discharged at 13th post operative day.

Figure 5: HE histology.
Histological analysis of the specimen revealed complete excision of the mass, with spindle cells without atypia, a low mitotic index, and positive immunohistochemical staining for CD99 and CD163, suggestive of AFH. The multidisciplinary group decided surveillance, with no further treatment, and at 6th year after surgery the patient has no symptoms or signs of recurrence or metastasis.

Figure 6: IHQ CD 163.

Figure 7: IHT CD 99.

DISCUSSION

AFH is a rare tumour first described by Enzinger in 1979. Initially believed to be a highly aggressive neoplasm with uncertain histogenesis, it primarily affects children and young adults, often involving superficial soft tissues of the extremities.

Although it presents distinct histological features, it remains challenging to diagnose due to its rarity, wide morphological spectrum, and lack of a specific immunohistochemical profile. AFH represents 0.3% of all soft tissue tumours and it is classified by the World Health Organization (2020) as a mesenchymal neoplasm of uncertain differentiation and intermediate biological behavior. It has a local recurrence rate of 15 to 20% and a distant metastasis rate of 1 to 3%.

While most cases are reported in children and young adults, our patient was older. The clinical signs in this patient were not related to mass effect, as reported in some literature, but rather to bleeding and subsequent hematoma formation caused by the lesion.

Before proceeding with the "complete" removal of the lesion, cytological examination of the lesion material was inconclusive but showed pigmented macrophages. The correct diagnosis was only made in the histology of the surgical specimen. Radiologic aspects were not conclusive but suggests persistent hematoma after 3 months of surveillance.

Immunohistochemical investigations are generally not diagnostic but are useful in supporting the diagnostic suspicion based on lesion morphology. Typically, markers such as smooth muscle actin (SMA), desmin, epithelial membrane antigen (EMA), CD68, and CD99 are positive. In particular, co-expression of EMA, SMA, and desmin is highly useful for diagnosis. In our patient it was positive for markers CD99 and CD 163. The range of lesions included in the differential diagnosis of AFH are several, from reactive lesions like granulomas to benign and malignant neoplasms. It is important to note that the only consistent finding in AFH is sheets of oval to spindle cells, while other features such as a fibrous capsule or lymphoplasmacytic infiltrate may be absent in the sample. Granulomatous inflammation may present with morphological characteristics similar to AFH, but it does not contain vascular spaces, unlike AFH.

Benign aneurysmal fibrous histiocytoma is another candidate in the differential diagnosis, but this entity is predominantly more heterogeneous than AFH. It occurs mostly in young adults, but is predominantly dermal and negative for desmin. Other candidates in the differential diagnosis are spindle cell hemangioma and/or Kaposi's sarcoma, which are positive for CD34 and human herpesvirus 8 (HHV8), unlike AFH. Another important aspect concerns the possibility of the development of this lesion in a patient who has previously undergone chemotherapy for prior renal cell carcinoma. Neither of these was the case of our patient.

CONCLUSION

AFH is a rare neoplasm with intermediate biological behavior, most commonly observed in the extremities of children and young adults. The vast majority of these neoplasms have an excellent prognosis, but correct diagnosis is important due to the risk of local recurrence and the small risk of metastasis and death. Lately, it has been identified in various unusual anatomical locations and has shown an increasing spectrum of morphological patterns, making its diagnosis challenging. In selected cases, it is important to perform molecular investigations.
or FISH to obtain a reliable diagnosis of AFH in order to reduce the risk of diagnostic error. Surgical excision and long-term clinical and radiological surveillance of these patients are highly recommended.

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