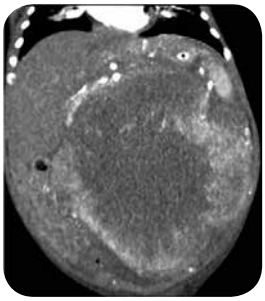

MYOFIBROBLASTIC TUMOR: A RARE ENTITY AND EVEN MORE RARE AGE OF PRESENTATION. A CASE REPORT

Tumor miofibroblástico: una entidad rara, a una edad de presentación neonatal aún más rara. Presentación de caso

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Palabras clave (DeCS)
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Summary

We present the case of a neonate, who upon physical examination a mass is palpated in the left hypochondrium, Abdominal ultrasound is performed with finding of a mass of probable adrenal origin; magnetic resonance imaging (MRI) shows a mass dependent on the mesentery. In surgery, important adhesion to neighboring organs is observed; biopsy is performed with the result of myofibroblastic tumor. This tumor is part of the heterogeneous group of mass-forming lesions called inflammatory pseudotumor, which is defined as a fibroblastic/myofibroblastic neoplasm, with intermediate biological potential, of greater occurrence in children. Given the low frequency of appearance in neonatal age, a review of the literature about its aetiology, diagnosis and treatment is carried out.

Resumen

Se expone el caso de un neonato, a quien durante el examen físico se le palpa una masa en el hipocondrio izquierdo. Le realizan ecografía abdominal con hallazgo de una masa de probable origen suprarrenal; la resonancia magnética (RM) muestra una masa dependiente del mesenterio. En cirugía se observa importante adhesión a los órganos vecinos, se le realiza biopsia con resultado de tumor miofibroblástico. Este tumor hace parte del grupo heterogéneo de lesiones formadoras de masa llamado pseudotumor inflamatorio, el cual se define como una neoplasia fibroblástica/miofibroblástica, con potencial biológico intermedio, de mayor ocurrencia en niños. Dada la baja frecuencia de aparición en edad neonatal, se realiza una revisión de la literatura acerca de su etiología, diagnóstico y tratamiento.

Introduction

Myofibroblastic tumor is part of the heterogeneous group called inflammatory pseudotumor, it is a true neoplasm given its clinical, pathological and molecular characteristics. According to WHO, it is a fibroblastic/myofibroblastic neoplasm with intermediate biological potential. It is predominantly located in visceral soft tissues, with local recurrence and metastases are rare (1). It is composed of myofibroblast cells in the spindle, accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils.

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Its etiology is not well de f ned, but may be associated with trauma or infections. The de f nitive diagnosis is histopathological, ALK (anaplastic lymphoma kinase) helps to establish the diagnosis, as images can not differentiate the in f ammatory pseudotumor from myo f broblastic tumor. The de f nitive treatment is complete surgical resection, although in some cases this entity has spontaneous regression, so there is discussion by some authors about the probable conservative management

Clinical case

Preterm newborn patient, product of twin pregnancy, delivery by caesarean section without complications, the physical examination is found mass in the left hypochondrium, abdominal ultrasound with f nding of mass that was initially suspected was of origin in the adrenal gland (Figure 1). Magnetic resonance imaging (MRI) is then performed to better characterize the lesion, resulting in mesentery-dependent mass (Figures 2, 3 and 4). In surgery, a mass with signi f cant adhesion to adjacent organs is found. For this reason, only biopsy is performed and pharmacological management is initiated. In the control tomography (CT) taken one month later, there is a signi f cant increase in the size of the mass that occupies practically the entire abdomen (Figure 5). The patient dies shortly after the control.

Discussion

Myo f broblastic tumor is an entity that is grouped within in f ammatory pseudotumors, is rare, especially in neonatal age and dif f cult to diagnose because of its clinical presentation, in images and its variable histological components (1-5).

Recently, association has been found with chromosomal abnormalities, such as translocations in the long arm of chromosome 2 and in the short arm of chromosome 9 (6) and cytogenetic aberrations, such as the ALK gene (anaplastic lymphoma kinase) (7, 8), p53 mutation and MDM2 expression, which could later be recognized as a speci f c neoplastic lesion or a true tumour (2, 3, 9-11).

Therefore, the most accepted theory of origin is an exaggerated in f ammatory response following localized trauma. Second, infection is proposed as antecedent, which is most often caused by mycobacteria, Epstein-Barr virus, Actinomyces and Mycoplasma. Other associations have been demonstrated with Mycobacterium avium-intracellular complex, Corynebacterium equi, Escherichia coli, Klebsiella, Bacillus sphaericus, Pseudomonas, Helicobacter pylori, Coxiella burneti and even HIV (6, 9, 12). Infectious aetiology is supported by the role of cytosines, mainly interleukin 6 (IL-6), for which a speci f c therapeutic approach has been described (9).

Myo f broblastic tumor can occur in any age group, but is most frequent in the f rst two decades of life with an average age at diagnosis of 10 years, with no signi f cant difference between the two sexes (1, 2, 13, 14). Tumor lesions are unique, but may be multiple in up to 5% of cases.

It manifests itself clinically with constitutional signs and symptoms depending on the site of origin and the effect of mass on neighboring organs, which are listed in Table 1 according to their frequency percentage.

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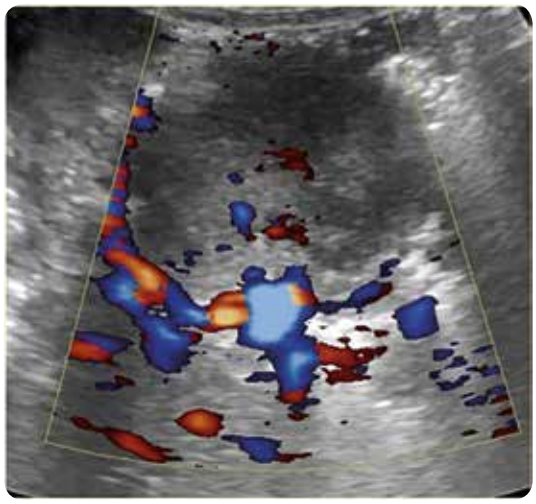

Figure 1. Ultrasound of the total abdomen, grey scale, mode B. Mass probably dependent on the left adrenal gland, solid aspect, heterogeneous echogenicity, defined contours, with flow of peripheral predominance after the application of color Doppler.

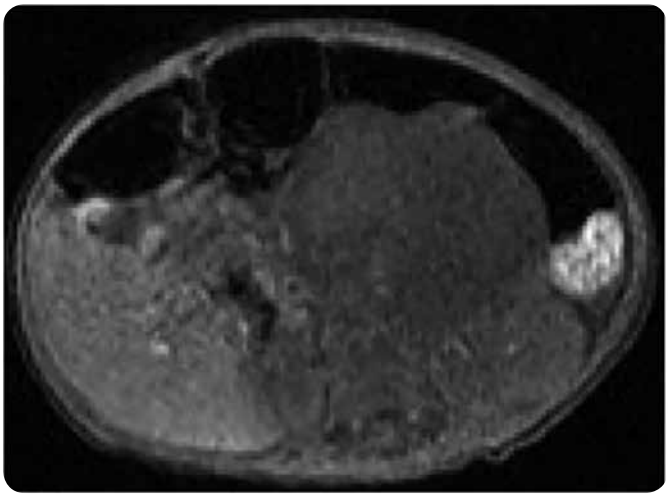

Figure 2. Abdominal MRI, sequence with simple axial T1 information: mass located in the mesentery, oval, low signal, and well-defined contours.

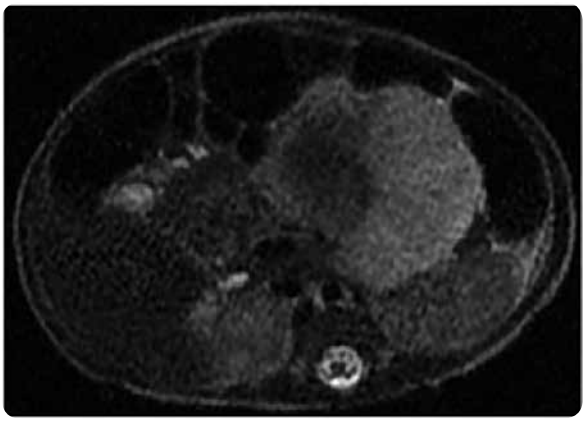

Figure 3. Abdominal MRI sequence with T2 axial information: oval mass located in the mesentery, well defined, predominantly high signal with areas of low signal due to probable necrosis.

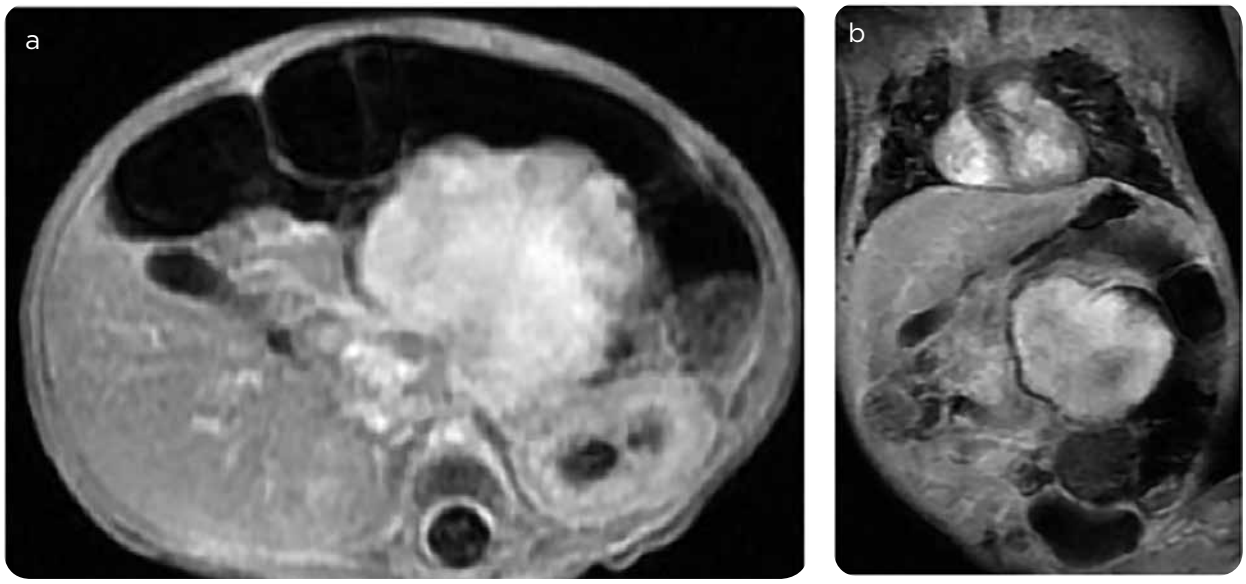

Figure 4. Abdominal MRI. a) Sequence with T1 information, axial, with contrast medium, b) Sequence with T1 information, coronal, with contrast medium. An oval mass with well-defined contours is identified, located in the mesentery, with important enhancement after the application of the contrast medium.

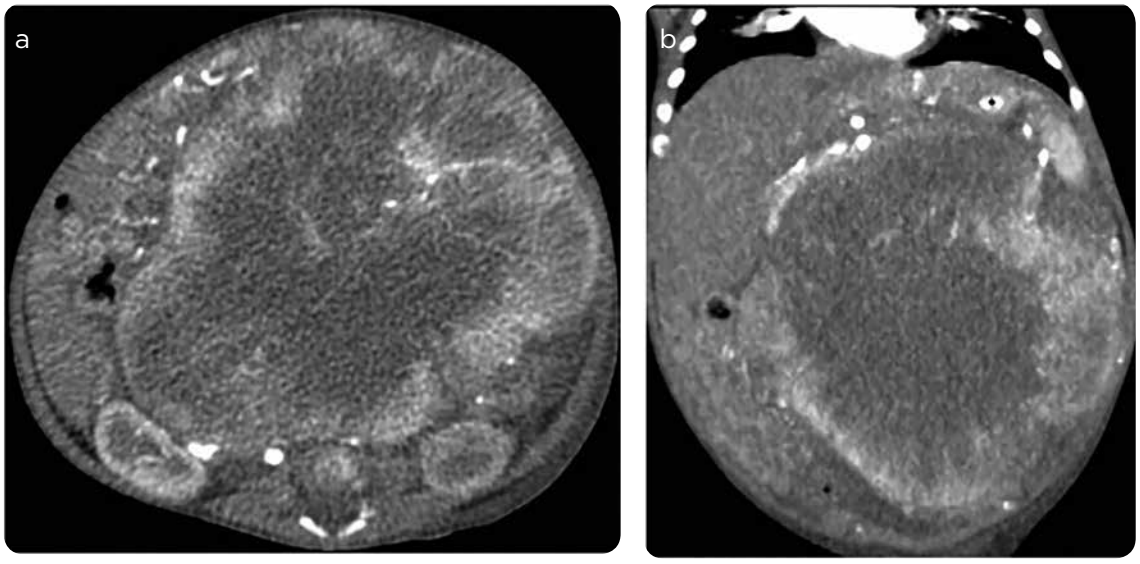

Figure 5. CT scan of abdomen with contrast medium, control one month after biopsy. a) Axial. b) Coronal. Mass with peripheral enhancement after administration of the contrast medium, with significant increase in size that occupies almost the entire abdomen, areas of low signal inside without enhancement of the contrast medium by necrosis, displacement of solid organs towards the abdominal periphery.

Table 1. Signs and Symptoms of Myofibroblastic Tumor

Systemic signs and symptoms	Porcentage
Abdominal pain	58,1
Fever	45,2
Weight loss	22,6
Loss of appetite	12,9
Nausea and vomiting	12,9

Source: Jun-Jie and others (3).

Laboratory results may indicate leukocytosis (34,6 %) and thrombocytosis, microcytic anaemia, abnormal hepatic function (30,8 %), C-reactive protein and high global sedimentation rate (19,2 %) which usually resolve after surgical resection.

Other findings are hypoalbuminemia, elevated tumour markers and anaemia. However, 15-40% of cases may be asymptomatic (1, 3, 4, 6).

On physical examination, in the majority of cases - as described here - a solid, palpable, unique mass is found, located anywhere in the abdominal cavity.

While MRI is the modality of choice for evaluating these lesions, the first-line imaging method is not considered. Abdominal ultrasound is considered the initial examination because of its high availability and low cost. In the exposed case, the additional use of color Doppler added useful diagnostic information (1).

In abdominal CT images the fibrous tissue is isodense to the muscle and after the application of the contrast medium a wide variety of enhancement patterns are observed, including late enhancement of the fibrotic component of the lesion, although in 10-25 % of cases it may present as a heterogeneous and calcified mass (1, 6).

In MRI images, in sequences with T1 information, fibrous tissue manifests with medium or low intensity and in sequences with T2 information the intensity is markedly low, with late and intense enhancement and with homogeneous or heterogeneous pattern (1, 15). However, in early stages, when the inflammatory pattern predominates more than the fibrous one, the sequence with T2 information can be observed with high signal. Another characteristic is that it does not show restriction to diffusion, this depends on the cellularity of the lesion.

The treatment of choice is complete surgical resection of the tumor with subsequent histopathological diagnosis; however, in the case exposed, due to the degree of invasion of neighboring organs only intraoperative biopsy was performed (3, 6). Other authors suggest conservative management using radiotherapy, chemotherapy, NSAIDs (COX-2 inhibitors) as adjuvant therapy, but there is no consensus about the effectiveness of these treatments which have been used for cases that are unresectable (6, 10, 16, 17). A new therapeutic option for myofibroblastic tumor is crizotinib, an ALK inhibitor (1).

When resection is not complete there may be recurrence usually during the first year (6,18). Recurrence cases are related to several factors including aneuploidy, which in 75% of cases presents malignant transformation (3), atypia and ganglion cells (19, 20).

Distant metastases are rare, occur in less than 5% of cases and have been associated with several factors, such as multifocality, intraperitoneal location and infiltration into adjacent structures. The most common sites of metastasis are lung, brain, liver and bone (1, 10).

Cases of spontaneous regression of the tumor have been reported, especially those located in the liver (3, 21-23).

Among the differential diagnoses in neonatal and infant age are, in order of frequency, inflammatory pseudotumor, infantile myofibromatosis, fibrosarcoma, granulomatosis, sarcoidosis, Langerhans cell histiocytosis, mesoblastic nephroma, Wilms tumor, polycystic kidneys, intestinal duplication cyst, mesenteric cyst, splenic cyst, neuroblastoma, lymphangioma and teratoma, among others (1, 23, 24).

Conclusions

Myofibroblastic tumor is an entity that is classified within inflammatory pseudo-tumors, can occur at any age, but is most common in the first two decades of life, with an average age at diagnosis of 10 years. Therefore, the exposed case of neonatal age is less frequent. According to WHO, it is a fibroblastic/myofibroblastic neoplasm with intermediate biological potential. It is predominantly located in visceral soft tissues and is difficult to diagnose, due to its variability in both clinical presentation and images. The treatment is the complete surgical resection of the tumour with subsequent histopathological diagnosis. When the resection is not complete, it can generally recur during the first year. Distant metastases are rare, occur in less than 5% of cases, the most common sites are lung, brain, liver and bone..

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