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Angiomyolipoma of the tunica dartos of the scrotum in infancy

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ABSTRACT

A 12-month-old boy presented with left scrotal swelling. The mass was irregular, soft, fleshy, and nontender. It adhered to the scrotal skin and gradually enlarged. Operative findings revealed a mass fixed to the scrotal fundus and diagnosed as angiomyolipoma. This is apparently the first report of scrotal angiomyolipoma in infancy.

Angiomyolipomas are benign mesenchymal tumors, first reported by Morgan et al. in 1951 [1]. Although it typically manifests as a renal mass, angiomyolipoma has been observed in numerous other organs [2–6]. These tumors are found mostly in the kidney and consist of three components: thick-walled blood vessels, smooth muscle cells, and variable amounts of mature adipose tissue. We report an angiomyolipoma of the tunica dartos of the scrotum in a 12-month-old boy that was diagnosed by a microscopic examination of a surgically resected specimen, and review the literature.

1. Case report

A 12-month-old boy presented with left scrotal swelling noticed for one month. The pregnancy was uncomplicated, with a normal vaginal delivery. There were no evident congenital abnormalities at birth. There was no familial history of diseases of the kidney and scrotum.

Physical examination revealed a 3 cm irregular, soft, and fleshy swelling in the left scrotum that was not tender when palpated. It adhered to the underlayer of the scrotal skin (Fig. 1). We doubted that the tumor was just a simple lipoma based on the physical examination. The cord structures appeared normal and the testis could be palpated separately from this swelling. The mass tended to increase slowly in size.

However, ultrasonography showed a single echogenic and hypovascular solid mass (Fig. 2). No other abnormalities were found on ultrasonography. The preoperative diagnosis was lipoma.

Operative findings revealed that the testicle and spermatic cord structures were normal. The mass consisted of yellow fatty tissue measuring 3 × 5 cm, appearing as a lipoma in a frozen section. The mass was fixed to the tunica dartos at the fundus of the scrotum, which appeared to be its origin (Fig. 3). The mass was completely resected with preservation of the testicle and spermatic cord structures. The scrotal skin at the site was secured, and the mass was fixed and preserved.

Examination of permanent sections of the lesion revealed mature fatty tissue with blood vessels and smooth muscle (Fig. 4a and b). On immunohistochemistry, vigorous staining of CD34 was demonstrated in the endothelial cells of the tumor (Fig. 4c). The final pathological diagnosis was suspected angiomyolipoma.

The postoperative course was uneventful. Six months after surgery, no gross disease was evident.

2. Discussion

Angiomyolipomas are benign mesenchymal tumors that arise mostly from the kidney. Extrarenal angiomyolipomas are rare, and most of them arise from the liver [7]. There are apparently no reports that discuss angiomyolipomas from the tunica dartos of the

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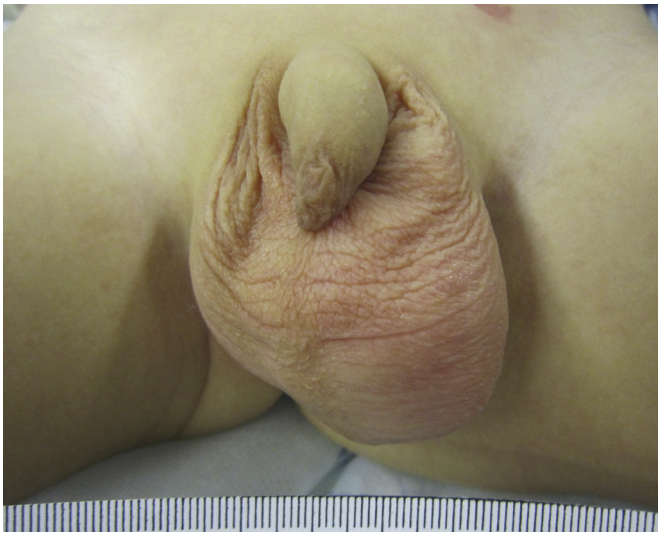

Fig. 1. Physical examination. Irregular, soft, fleshy mass, nontender swelling in the left scrotum.

scrotum in infancy. Although an angiomyolipoma is usually a noninvasive tumor that does not metastasize, its capacity to cause hemorrhage and other significant clinical symptoms, as well as its characteristic cellular composition and distinctive molecular biology, have made it the subject of continuing interest among researchers and clinicians [8].

An angiomyolipoma is commonly described as a hamartoma, meaning the abnormal proliferation of tissues that are normally present in the organ, although it has been argued that it is actually a choristoma, since fat and smooth muscle are not normal kidney components [8,9]. However, there are strong medical data supporting its status as a "neoplasm." Green et al. [10] demonstrated that the X chromosome is inactive in angiomyolipoma cases that developed in tuberous sclerosis patients and thus, this neoplasm has clonal origins. There are two types of renal angiomyolipoma which can be differentiated clinically. The first type is seen in tuberous sclerosis patients, develops bilaterally, and could have multiple centers. Angiomyolipoma develops in 80% of tuberous sclerosis cases. The second type is unilateral. However, neither type can be histologically differentiated [11–13].

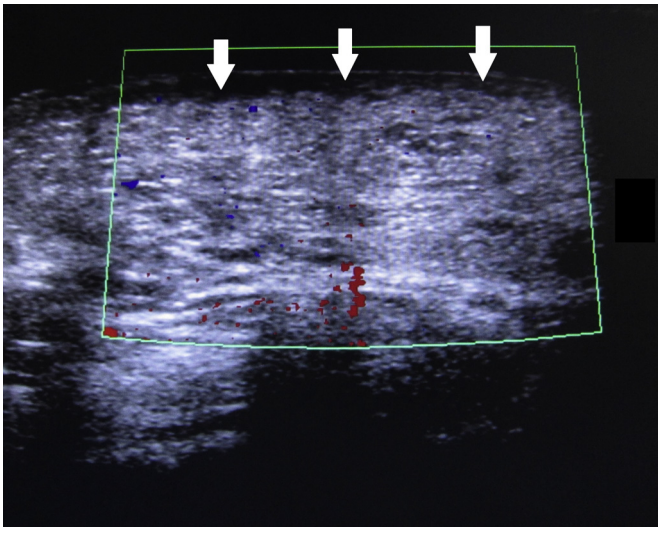

Fig. 2. Ultrasonographic image of the left lower abdomen. Single echogenic and hypovascular solid mass (*arrow*).

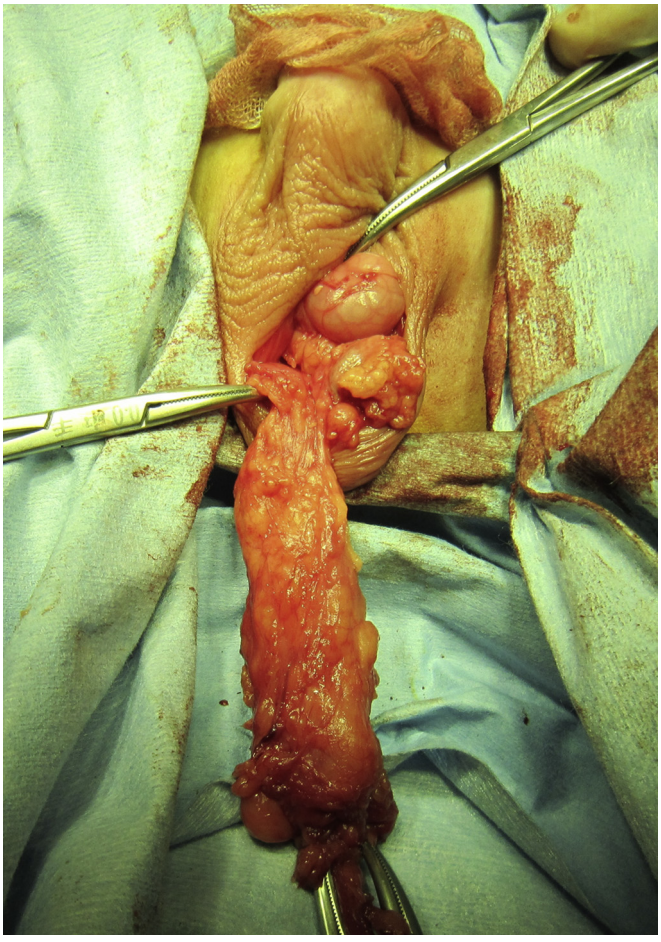

Fig. 3. Operative findings. The yellow fatty tissue was fixed to the tunica dartos at the fundus of the scrotum.

The most frequent symptoms in renal angiomyolipoma patients are acute flank pain, hematuria, and a palpable mass [14]. Solitary tumors of renal angiomyolipoma may cause an acute abdomen and shock as a result of spontaneous hemorrhage in the tumor. Yamakado et al. [15] reported that a tumor of 4 cm or larger was a predictor of spontaneous rupture. In our case, there were no symptoms. However, although the mass was mostly soft and fleshy, there was also a partially indurated region that was not tender when palpated. Therefore, we doubted that this tumor was just a simple lipoma.

Angiomyolipomas can usually be diagnosed using ultrasound and computed tomography (CT) imaging [8]. The ultrasonographic appearance of angiomyolipoma is significant for its marked hyperechoic signal and acoustic shadowing. However, there was no hyperechoic lesion in our case. Since the tumor did not have a sufficiently large muscular component or intratumoral hemorrhage, the lesion could not be categorized by ultrasonography [16,17]. On the other hand, the characterization of angiomyolipoma by CT depends on identifying fat in the lesions. In 4.5% of angiomyolipomas, no fatty elements can be detected on CT scans and these fatty elements can be detected only by microscopy [18,19]. This type of angiomyolipoma is called an angiomyolipoma with minimal fat. Because of this limitation, an accurate diagnosis by radiological findings alone is difficult. Therefore, we thought that ultrasonography would be more beneficial than CT preoperatively.

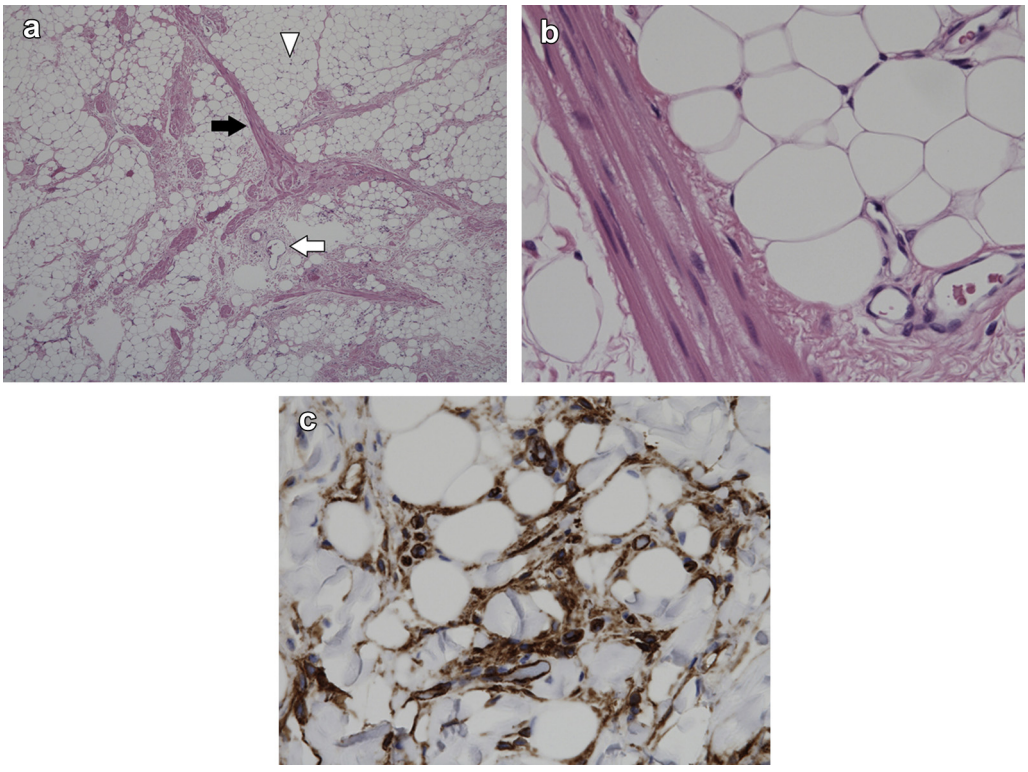

Fig. 4. Examination of permanent sections. a) Histopathology demonstrated concurrent presence of fat (*arrowhead*), vascular structures (*white arrow*), and smooth muscle (*black arrow*) [Hematoxylin & Eosin, ×10]. b) The tumor consisted of spindle cells, adipose tissue, and blood vessels of several sizes [Hematoxylin & Eosin, ×400]. c) Positive staining for CD-34 [×400].

The indications for treating asymptomatic angiomyolipoma are controversial. As a benign lesion that is usually asymptomatic, angiomyolipoma may often not require intervention [8]. Indications for intervention include suspicion of malignancy, spontaneous hemorrhage causing significant symptoms, pain, hematuria, or risk of rupture or other complications. For these reasons, attempts have been made to manage most lesions conservatively. Repeat ultrasound yearly or semi-yearly can be performed to evaluate disease stability versus progression.

The treatments of renal angiomyolipoma are angiographic embolization, cryotherapy, and tumor resection [8]. Since our case was a child with a scrotal tumor, we thought it would be inadvisable to perform embolization or cryotherapy. Furthermore, we thought that an obvious advantage of surgery would be that a pathological diagnosis could be made. We therefore performed surgical resection.

The histological diagnosis of angiomyolipoma is usually easy because of its distinctive and pathognomonic histological findings. The gross sections of the tumors are mostly yellow because of their fat content. Angiomyolipomas are composed of mature fat cells, vascular tissue, and smooth muscle. The fatty component is typically normal in appearance. The vascular components are often tortuous and thick walled, have a lower elastin content than normal vessels, and are commonly surrounded by a cuff of smooth muscle cells [20]. The absence of elastic tissue in the vessels of tumors predisposes the patient to the formation of an aneurysm and spontaneous hemorrhage. Smooth muscle, which appears as normal spindle cells or rounded epithelioid cells, is present in varying amounts, from focal areas near blood vessels to predominant sheets comprising the majority of the tumor. Smooth muscle nuclei are generally small and regular, although some areas occasionally have enlarged atypical nuclei with nucleoli and rare mitotic figures [8,21]. On immunochemistry, the melanosome-associated protein HMB-45 is consistently expressed in angiomyolipomas [8]. Other markers that are commonly positive in angiomyolipomas include smooth muscle actin, CD-34, and CD-68 [8,22]. Kawaguchi et al. [23] reported that the immunohistochemical findings for smooth muscle actin were positive in 30% of typical angiomyolipomas, but that they were negative in the malignant type.

In general, an angiomyolipoma is a tumor with a benign clinical course [8]. However, an incomplete excision may result in local recurrence. Moreover, there have been several reports of angiomyolipomas with high malignant potential, known as epithelioid angiomyolipomas [23,24]. These tumors have the potential for local invasion and distant metastases, but the involvement of regional lymph nodes by these tumors is uncommon [8]. There was no evidence of local recurrence of angiomyolipoma in our case. However, we think that this should be followed up regularly.

3. Conclusions

This is apparently the first case of angiomyolipoma of the tunica dartos of the scrotum reported in infancy. Although extremely rare, it should be considered in cases of scrotal tumor.

Conflict of interest

There are no declared conflicts of interest.

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