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Asymmetric labium majus enlargement in an adult: A case report

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ABSTRACT

A 37-year-old woman presented with a right labium majus thickening with no palpable mass, which had been evolving for a few months. A partial right labium majus ablation was performed at the request of the patient for esthetic reasons. The lesion was most histologically similar to CALME (childhood asymmetric labium majus enlargement). The lesion was benign, but several other differential diagnoses were considered. Two years after the surgery, there had been no recurrence. To our best knowledge, this is the first such case reported in an adult.

1. Introduction

The present paper discusses the case of a 37-year-old woman with a labium majus thickening with no palpable mass. Based on its histological characteristics, the lesion was similar to CALME (childhood asymmetric labium majus enlargement), first described in 2005 by Vargas et al.: poorly circumscribed with normal vulvar tissue, and containing characteristic fibroblastic cells, adipose tissue, blood vessels and abundant nerves [1,2]. The condition is unusual in an adult female. Several differential diagnoses are proposed as well as the imaging particularities and treatment options. It seems that our case is the first described in an adult.

2. Case Presentation

2.1. Clinical Findings

A 37-year-old woman with no significant medical history presented with a right labium majus thickening, which had been evolving for a few months. There was no palpable distinctive mass (Fig. 1).

During the one-year follow-up, the lesion appeared stable, although the patient reported a reduction in its size after restricting her cycling to less than one hour per day. There was no associated bleeding or pain.

2.2. Diagnostic Assessment

An ultrasound scan showed a hypoechoic mass measuring 40x38x13mm – a fatty, circumscribed and vascularized lesion, on color Doppler resembling an atypical lipoma with degeneration. At this stage, liposarcoma could not be ruled out.

Magnetic resonance imaging (MRI) showed enlarged labial tissue composed of contrast-enhanced hypointense signal on T1-weighted imaging and moderately hyperintense signal on T2-weighted imaging (Fig. 2).

2.3. Therapeutic Interventions

The patient asked for a surgical removal for esthetic reasons. Intraoperatively, surgeons observed apparently normal tissue with skin thickening. No lipoma structure or lymphatic dilatation was found. A partial right labium majus ablation was performed.

The pathology report indicated an asymmetric labium majus enlargement with no evidence of tumoral proliferation. The lesion consisted of normal vulvar soft tissue with a predominance of a fibrous component (Fig. 3). The lesion was most histologically similar to CALME: poorly circumscribed with normal vulvar tissue, and containing characteristic fibroblastic cells, adipose tissue, blood vessels and abundant nerves [1,2]. Such presentation has not formerly been described in an adult.

2.4. Follow-up and Outcome

Six months after the surgery, the good cicatrization of the operative site was noted, no recurrence was found, and the patient was completely satisfied. The follow-up within two years was proposed.

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Fig. 1. Right labium majus thickening, first clinical presentation.

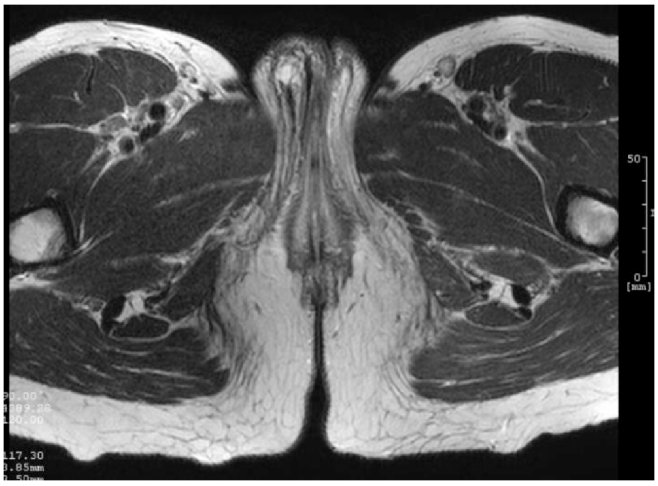

Fig. 2. MR T2-weighted imaging showing a hyperintense signal.

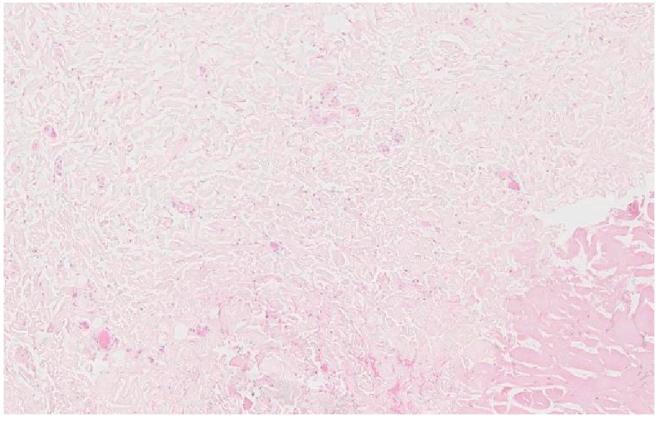

Fig. 3. Hematoxylin eosin x100: non-tumoral vulvar stroma with delicate collagen bundles (lower right).

3. Discussion

The pathology report indicated normal vulvar soft tissue with a predominance of the fibrous component and no evidence of tumoral proliferation. This description aligns with the histological findings in CALME, first described in 2005 by Vargas et al., previously known as hamartoma, fibroma, fibrous hyperplasia or lipoma [1]. As of today, there has not been a report of this histological presentation in an adult.

Our MR images partly differ from other publications: we observed a contrast-enhanced hypointense signal on T1-weighted imaging and a moderately hyperintense signal on T2-weighted imaging, while Salvatori et al. described the hypointense signal on T1 but a hypo-isointense signal on T2-weighted imaging [3]. Similarly, Gokli et al. reported hyperintense linear septations on T2 [2], whereas Vargas et al. described four patients with hypointense signal on T2, one patient with isointense signal on T2, and finally three patients with hyperintense signal on T2-weighted imaging [1].

The lesion was benign, but several differential diagnoses were considered, as described by Salvatori et al., such as: mimicking lesions; inguinal hernia (which is distinguished by the fluctuating nature); vascular malformations (bluish discoloration of the skin and history of pain and thrombosis); lipoma and neurofibroma (both well-defined masses on palpation); Bartholin's duct cyst or abscess (pain and erythema); labial hypertrophy (bilateral); rhabdomyosarcoma; and angiofibroblastoma [3]. Additional differential diagnoses were proposed by different authors, such as: Crohn's disease of the vulva; perineal nodular induration and aggressive angiomyxoma; vulvar metastasis; and finally infectious etiologies such as candidiasis, schistosomiasis and enterobiasis [2,4–6]. McCluggage and colleagues had described a unilateral pseudoneoplasic lesion on the vulva occurring in competitive female cyclists; they term this lesion as reactive fibroblastic and myofibroblastic proliferation of the vulva or "cyclist's nodule" [7]. In our case, the patient was not a competitive cyclist, although she reported a reduction in size after restricting her cycling. There was no myofibroblastic proliferation and less hyalinization according to our pathology report.

Generally, diagnosis is based on histological evaluation by biopsy or surgical excision [2,4,5]. According to Vargas et al., the most histologically similar lesion is aggressive angiomyxoma [1]. Like CALME, aggressive angiomyxoma is a poorly circumscribed vulvar lesion, characterized by paucicellularity with fibroblastic cells and abundant extracellular myxoid and a moderately collagenized matrix, the stroma being light eosinophilic with numerous blood vessels [1,8]. The major difference between CALME and aggressive angiomyxoma is the soft-tissue infiltration characterized by entrapment of muscle, nerve and adipose tissue [8].

A conservative approach with close clinical and sonographic follow-up is preferred - biopsy and/or excision being reserved for atypical cases or cosmetic reasons [2]. Although a surgical excision may lead to a recurrence in 50% of cases [1,3], it was neither observed with our patient at the first six-month follow-up, nor with the two cases described by Salvatori et al. [3].

4. Conclusion

After the surgical removal, the pathology report demonstrated normal vulvar soft tissue with a predominance of a fibrous component and there was no evidence of tumoral proliferation or infiltration. The first clinical and radiological hypothesis of degenerated lipoma was ruled out. The lesion was most histologically similar to CALME. At the six-month follow-up there was no recurrence observed. It seems that our case is the first ever described in an adult.

Contributors

Elze Prokurotaite is the lead author, and was responsible for conception and design of the manuscript, critical revision of the article for important intellectual content, and final approval of the version to be submitted.

Nicolas Sirtaine contributed to analysis and interpretation of data, and revising the article critically for important intellectual content.

Frédéric Buxant contributed to analysis and interpretation of data,

and final approval of the version to be submitted, and was involved in patient care.

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Patient consent

Obtained (the patient consent to the publication of the report and any accompanying images has been given).

Provenance and peer review

This article was not commissioned and was peer reviewed.

Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this case report.

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