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Case report

Abdominal wall endometriosis misdiagnosed as a desmoid tumor: A case report

Martina Girardi [a], Alessandra Marano [a], Mirella Fortunato [b], Enrico Gelarda [a], Maria Carmela Giuffrida [a,*]

[a] Department of Surgery, General and Oncologic Surgery Unit, Santa Croce e Carle Hospital, Via M. Coppino 26, 12100 Cuneo, Italy
[b] Department of Pathology, Santa Croce e Carle Hospital, Via M. Coppino 26, 12100 Cuneo, Italy

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ABSTRACT

Introduction: Abdominal wall masses have different aetiologies. Diagnosis includes desmoid tumors (DTs) and other benign and malignant lesions, among which abdominal wall endometriosis (AWE). Diagnosis is challenging if symptoms are aspecific, and the contribution of imaging may be weak. We present a case of AWE that according to clinical history and imaging was misdiagnosed as DT.

Presentation of case: A healthy 35-year-old female presented, 4 years after a cesarean delivery, a rapidly growing painless subumbilical mass within the right rectus abdominis muscle. Ultrasound and magnetic resonance imaging suspected a DT. The patient underwent complete resection of the mass and pathological examination revealed foci of endometriosis in the muscle. Patient's post-operative course was uneventful and at 18-month follow-up, no recurrence has been detected.

Discussion: The current case highlights differences in clinical presentation and imaging in case of AWE and DTs, underlining possible pitfalls in diagnosis. In young women with previous gynaecological abdominal surgery, AWE is the most likely disease when a mass in the region of the scar appears. Differential diagnosis is complex and rare entities like DTs should nevertheless be taken into consideration. A complete surgical resection with negative margins is considered the primary treatment for AWE and for selected DTs. Final pathology of the tumor can state the precise diagnosis.

Conclusion: Since AWE and DTs share similar clinical signs and aspecific imaging exams, both diseases should be considered in case of abdominal wall mass in female patients of childbearing age and history of uterine-related surgery.

1. Introduction

Differential diagnosis of abdominal wall masses (AWMs) includes benign and malignant tumors, haematomas, inflammatory/fibrotic lesions, and endometriosis. Clinical history and imaging allow categorization of lesions into subtypes and guide management. In females of reproductive age presenting an anterior AWM, desmoid tumor (DT) remains one of the most likely diagnoses [1]. Nevertheless, abdominal wall endometriosis (AWE), a disease mainly of gynaecological interest, can be another common cause of AWM in this clinical setting.

DTs, also known as aggressive fibromatoses, are rare and locally aggressive fibroblastic proliferations of fibrous tissue that can develop in any muscular aponeurotic structure of the body [2]. Sporadic abdominal DT usually arises in the rectus abdominis muscle and presents as a solitary, painless slow-growing mass. A typical patient with abdominal wall DT is a female of reproductive age, usually having previously given birth [3].

AWE is defined by the finding of any ectopic endometrium between the parietal peritoneum and the skin and generally develops within or adjacent to a surgical scar [4]. Patients are typically young women with a history of previous obstetric or gynaecological surgery, presenting with tenderness and swelling of the abdominal wall scar.

We report a case treated in a tertiary referral center of a young woman with an AWM, clinically and radiologically diagnosed as a DT, while pathology revealed as AWE. We underline the difficulties of a correct diagnosis of an AWM in case of atypical presentation.

The case was reported according to the Surgical CAse Report (SCARE) criteria [5].

* Corresponding author at: Department of Surgery, General and Oncologic Surgery Unit, ASO Santa Croce e Carle, Via M. Coppino 26, 12100 Cuneo, Italy.
E-mail address: giuffrida_mc@ospedale.cuneo.it (M.C. Giuffrida).

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2. Presentation of case

A healthy 35-year-old female with BMI 26.2 presented complaining of a 4-month history of a painless mass in the right inferior abdominal quadrant, rapidly increasing in size in the last two months. No pelvic pain or history of symptomatic endometriosis was referred. The patient had undergone a Cesarean section 4 years before the initial presentation. Physical examination revealed a 3-cm length muscle swelling in the context of the right abdominal wall, located cephalad and lateral to the Pfannestiel scar.

Blood chemistry was regular. Ultrasound (US) evaluation showed a 25 mm long fusiform inhomogeneous structure in the right rectus abdominis sheath. Doppler US flow evidenced blood vessels within the lesion (Fig. 1). Magnetic resonance imaging (MRI) confirmed a mass characterized by isointensity in the axial unenhanced T1-weighted images, light signal hyperintensity in the T2-weighted sequences, and conspicuous enhancement after administration of a gadolinium contrast agent. The lesion presented feeding vessels and irregular margins (Fig. 2). MRI images did not reveal any association to the surgical Pfannestiel scar, and no intra-abdominal lesions were found. Overall, the finding was not of univocal interpretation, but suspected of neoplastic lesion of mesenchymal series.

The case was discussed during our multidisciplinary consultation. Because of the rapid growth of the mass, which nevertheless appeared easily resectable, surgical excision was preferred over biopsy. The procedure was performed by a single surgeon (MC.G.) with extensive experience in abdominal cancer treatment.

2.1. Operative technique

Thromboprophylaxis and cephazoline 2 g i.v. were administered 12 h and 1 h before the surgery, respectively. Following general anaesthesia, the patient was placed in a lithotomy position. A lozenge 8 cm-length incision was conducted along the right paramedian line. The fascial plane was then identified, and the lesion and its margins were determined in the context of the rectus muscle with the guidance of an intraoperative US. A complete excision was conducted down to peritoneum, that did not appear infiltrated by the lesion. Peritoneum and fascia were easily closed in a running fashion with absorbable sutures. A spiral drain was placed at the sub-fascial level.

Total operative time was 80 min. After an uneventful postoperative course, the patient was discharged on post-operative day 2.

2.2. Pathologic findings

On gross examination a fragment of abdominal wall was observed consisting of skin and subcutaneous muscle with a single, firm, whitish nodule in correspondence of the muscle tissue with fine, yellowish-red dots and irregular margins having a maximum diameter of 13 mm. Microscopic examination of the specimen showed a lesion that, stained with haematoxylin and eosin, revealed fibrous areas englobing endometrial glands lined by pseudocolomnar epithelium, with bland nuclear morphology within a decidualized stroma.

Immunohistochemical staining of formalin-fixed, paraffin-embedded sections was performed. The endometrial-type glands and stroma were positive for estrogen receptors. CD10 immunostaining highlighted endometrial stromal cells.

A pathological diagnosis of endometriosis is made upon the identification of the least two of three key elements, as occurred in the present case i.e. endometrial stromal and epithelial cells and signs of chronic bleeding in or adjacent to endometrium-like tissue. Fibrosis is commonly observed surrounding endometriotic implants and possibly represents extensive inflammation and tissue remodeling.

Based on these microscopic findings and immunohistochemical staining results, the nodule was diagnosed as endometriotic foci (Fig. 3).

Given the R0 exeresis of the mass, a follow up program was proposed. At 18-month follow-up, the patient remains free of symptoms and no recurrence has been detected.

3. Discussion

Here we presented the case of a resectable AWM, clinically diagnosed as a mesenchymal series tumor, most probably a DT, and revealed at the pathological examination as an extra-pelvic endometriosis. A complete surgical removal warranted acceptable outcomes.

DTs are rare, unique mesenchymal neoplasms, with no metastatic potential, but a high tendency to recur locally after excision, even after complete surgical resection [6–8]. They most often occur sporadically (90%) [8] and represent the most common AWMs in young women without a relevant medical history. Supposed risk factors of DTs are previous surgical interventions, pregnancy, and hormonal treatment with oestrogens [9]. In the case presented, the local tissue trauma of a cesarean section in the clinical history of the patient could have been a possible risk factor for the onset of the disease. DTs can develop at any body site mostly presenting as a deeply seated painless or minimally painful mass with a history of slow growth.

AWE is the most common type of extra pelvic endometriosis, and it is mostly associated with a history of obstetric or gynaecological procedures. This disease, usually within proximity to a prior surgical incision [10], has a rare occurrence and its classic symptomatology consists of an intermittently painful mass that is very sensitive to palpation and increases in volume and sensitivity according to the phase of the menstrual cycle. However, this classic history is only present in approximately 50% of patients with AWE [11].

In the current case, even though the patient was of reproductive age with a cesarean section history, she did not complain of abdominal pain,

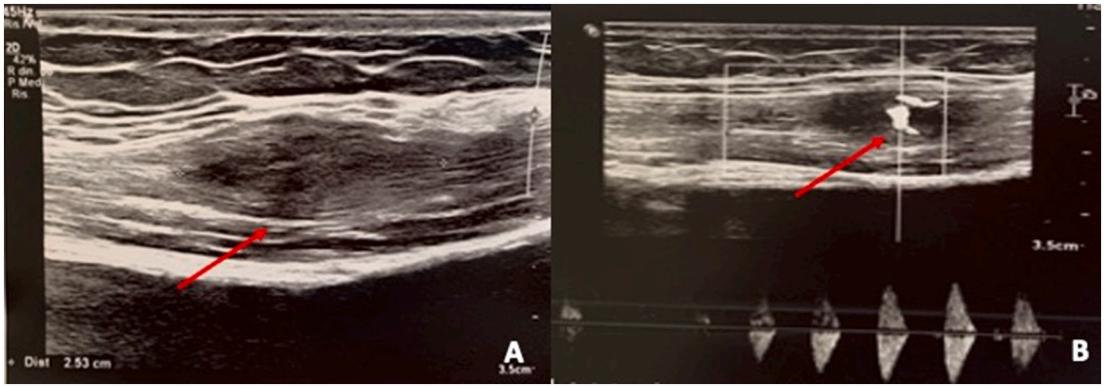

Fig. 1. Ultrasound preoperative image showing a 25-mm hypovascular hypoechoic mass in the inferior right rectus abdominis muscle (arrow, A) and doppler US flow evidencing blood vessels within the lesion (arrow, B).

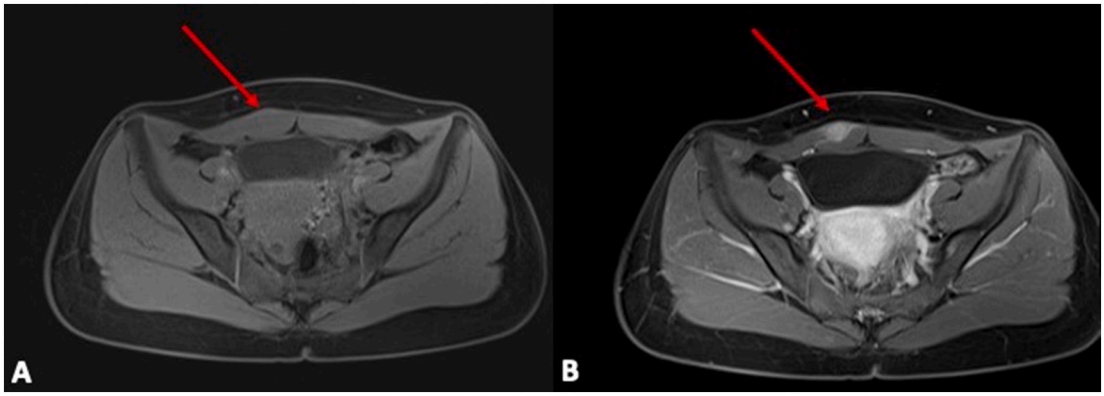

Fig. 2. Axial unenhanced T1- weighted MR images showing a hypointense mass within the right rectus abdominis (arrow, A). After administration of a gadolinium contrast agent, there was progressive homogeneous enhancement of the mass (arrow, B).

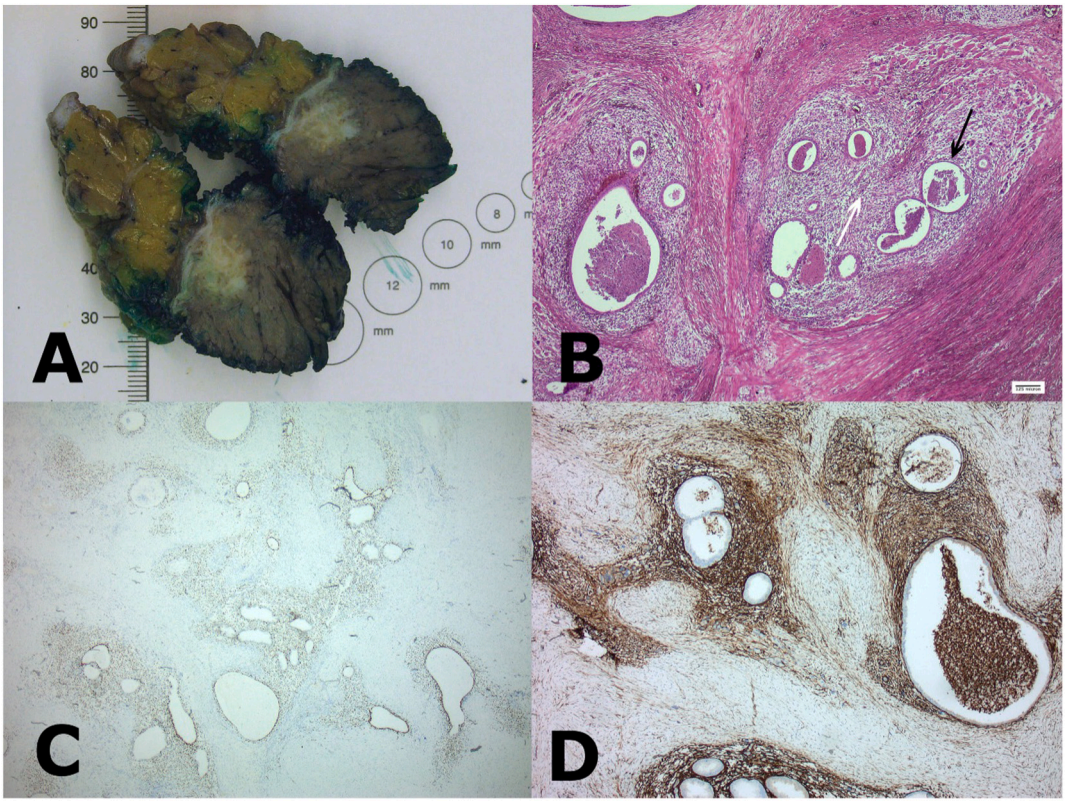

Fig. 3. Pathology.
On gross pathologic examination the fragment of abdominal wall evidenced a whitish nodule in the subcutaneous muscle (A). Microscopic pictures with haematoxylin and eosin staining show nodule with extensive fibrosis and tissue remodeling surrounding islands of endometrial-type glands (black arrow) and stroma (white arrow) with foci of bleeding and macrophages containing blood pigment. Magnification 4× (B). Immunohistochemical staining for estrogen receptors evidenced nuclear stains in the stroma and epithelial cells. Magnification 2× (C). CD10 shows strong cytoplasmic staining of the stromal cells. Magnification 4× (D).

and she did not experience significant catamenial changes in the mass. For these reasons, DT was considered the most likely diagnosis. However, AWE is often not directly below the surgical scar since the fascial incision often extends more laterally and superior to the skin incision [4].

DTs and AWE have several similarities also regards radiological imaging. For DTs, there are no pathognomonic sonographic features [12], but the most common characteristics are the oval shape of the lesion, well or poorly defined margins and variable echogenicity. Doppler pattern suggests the presence of blood vessels within the tumor. Similarly, AWE appears in US as a heterogeneous hypoechoic, solid mass. Margins are irregular, often spiculated and may appear to infiltrate adjacent tissues. US evaluation can guide the diagnosis by locating the tumor: an intramuscular or aponeurotic site indicates mostly a DT, whilst a subcutaneous topography may be suggestive for an AWE [13].

Concerning DTs, MRI characteristics are variable with hypo- to isointensity at T1-weighted images and hyperintensity at T2-weighted images in the earlier stages [11]. DTs typically show moderate to marked contrast material enhancement.

MRI features of AWE are usually heterogeneous on both T1 and T2-weighted images and may show contrast enhancement [1]. An MRI aspect that is highly suggestive for parietal endometriosis is a well-delimited subcutaneous solid mass with the infiltration of the

muscular aponeurosis [2].

Imaging in our patient was consistent with the diagnosis of DT, due to the intramuscular location of a lesion quite distant from the surgical scar, its spindle shape, and the US and MR features.

In case of AWMs, biopsy may be performed for suspicious masses to confirm the diagnosis but may not be necessary if complete R0 resection can be achieved [8]. In the current case, since it was a small and resectable lesion, a preoperative biopsy was not performed, and the complete excision of the lesion was chosen for diagnosis and definitive treatment.

Surgery is the primary treatment for patients with resectable DTs [8]. Negative microscopic margins resection should be the goal, but positive microscopic margins can be accepted when function or cosmesis is an issue [8,14]. However, recent updated guidelines have included observation as an option for selected patients with resectable DTs [8] under supervision of an experienced team [3,14].

Likewise, a R0 resection without mass rupture is considered the primary treatment for AWE. The excision must be wide enough to remove all skin segments, subcutaneous tissue, muscles, aponeuroses, and the peritoneum potentially involved. If the fascia defect is big, the insertion of a mesh may be required [15]. In the current case, a wide excision was carried out with negative margins down to the peritoneum and the closure was performed without a mesh due to the small size of the fascia defect.

4. Conclusion

In the presence of AWM in reproductive women, DTs and AWE can be misdiagnosed since the clinical signs are similar, and the imaging exams are aspecific. Surgeons should maintain a high suspicion for AWE in women of childbearing age with a palpable abdominal mass and history of uterine- relating surgery.

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Ethical approval

This study was exempted from ethical approval at our institution, however, permission was sought from the patient in question to present and publish this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

Martina Girardi: Conceptualization, Writing- original draft, Data Curation.

Alessandra Marano: Methodology, Writing - review and editing, Supervision.

Enrico Gelarda: Resources.

Mirella Fortunato: Resources.

Maria Carmela Giuffrida: Conceptualization, Writing- review and editing, Final revision.

Registration of research studies

None.

Guarantor

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Declaration of competing interest

None.

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