ATYPICAL SYMPLASTIC LEIOMYOMA, UNUSUAL HISTOPATHOLOGICAL VARIANT OF LEIOMYOMA - CASE REPORT

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Introduction

We present a case of atypical leiomyoma presented with abdominal mass and bowel symptoms. This is a rare type of leiomyoma called atypical symplastic leiomyoma. Uterine leiomyomas are the commonest gynaecological benign disease. Rare variants with distinct histological and molecular features have been described and these include mitotically active leiomyoma, cellular leiomyoma, atypical symplastic leiomyoma and smooth muscle tumours of uncertain malignant potential (STUMP). These pose a diagnostic dilemma as they have some features suggestive of malignancy.

Case Report

A 43-year-old Caucasian woman was seen in the Gynaecology rapid access clinic following referral for a large pelvic mass. She had first presented to the Accident and emergency (A&E) department with a 6-day history of abdominal pain, nausea and abdominal distension. She had no urinary symptoms. An X-ray was performed and excluded subacute intestinal obstruction but had shown a large central area of increased opacification projected over the sacrum and L2-L4 vertebra. A follow up CT scan showed massive uterine enlargement with centrally placed heterogenous density lesion 16.5 X 9.5 X 13.5cm suggestive of either a massive submucosal fibroid, hematometra or malignant neoplasm. (Figure 2). She had never been pregnant, was not on any contraception and had regular but heavy periods. She suffers from osteoarthritis for which she occasionally takes simple analgesia and has previously undergone partial left medial meniscectomy. Her BMI is 47Kg/m^2. She has bilateral lymphoedema. Transabdominal ultrasound scan showed a large midline abdominopelvic mixed heterogenous/hypoechoic mass 21 X17 X14cm in size. (Figure 1) Haematological investigations were all normal with a haemoglobin count of 119g/L and CA 125 of 24u/ml.

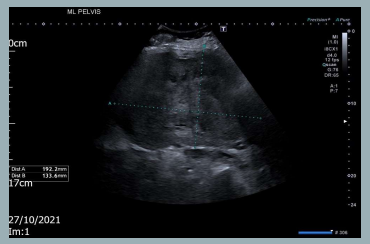


Fig 1

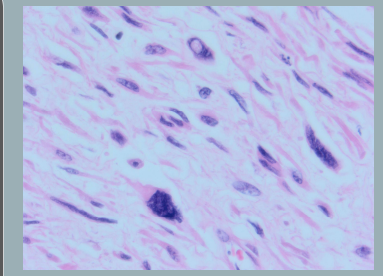


Fig 4

Method

The patient had total abdominal hysterectomy and bilateral salpingo-oophrectomy. Intra operative findings were of a large uterine mass potentially a fibroid, adenomyoma or sarcoma. (Figure 3)

Both tubes and ovaries were adherent to the pelvic wall but grossly normal. She made an uneventful recover.

Histological diagnosis of atypical symplastic leiomyoma was made due to the presence of a focal area with bizarrely shaped, hyperchromatic nuclei and no mitotic figures or tumour necrosis. (Figure 4)

Conclusion

Affecting 20-30% of women aged between 30 and 40, leiomyoma are common gynaecological occurrence. Some leiomyomas have an atypical histological appearance which might make it problematic to differentiate from leiomyosarcoma. Histopathological differentiation can be difficult. However, accurate diagnosis is important to counsel the patient and to plan appropriate follow up.



Fig 3



Fig 2

Discussion

Leiomyomas are the most common uterine neoplasm and the often originate from the myometrium. The gold standard for management is surgical hysterectomy or myomectomy. Histological examination is important to rule out uterine sarcoma. On histology, they predominantly contain smooth muscle cells with variable amounts of connective tissue. The absence of mitotic features in leiomyomas is key and differentiates leiomyomas from malignant leiomyosarcomas. Taylor and Norris introduced the term atypical leiomyomas in 1966 in reference to leiomyomas with cytologic atypia. These were further characterised in 1994 by Bells criteria as containing a triad of smooth muscle cells with moderate to severe cytological atypia, absent tumour necrosis factor and fewer than 10 mitotic figures per high power film mitotic index. The uterus is the commonest site of occurrence of atypical leiomyomas although they have also been reported in other organs such as the scrotum, vagina and nasal cavity. Presentation and clinical findings are in most cases like typical leiomyomas. In extremely rare cases, leiomyomas can transform to leiomyosarcomas, and patients have a poorer prognosis. The reported incidence of leiomyosarcoma in uterine leiomyomas is estimated to be between 0.13 to 0.29%. The rate of transformation has not been found to be increased in the atypical variants. Following histology, immunohistochemical examination of oestrogen receptors (ER) progesterone receptors (PR) and the makers Ki67 and p53 are useful for further differentiation in cases of uncertainties. The expected profiles for mean expression are as shown in table 1

	Immunohistochemical Marker (%)			
Diagnosis	ER	PR	p53	Ki67
Leiomyoma	63	75	0.3	0.6
Symplastic leiomyoma	85	89	0.6	5.5
Leiomyosarcoma	38	39	52	61

Table 1