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Dermatofibrosarcoma Protuberans in pregnancy: A rare occurrence

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Abstract
Background: Derma to fibro sarcomaprotuberans (DFSP) is an unusual fibro histiocytictumour classified as an intermediate-grade malignancy characterized by infiltrative growth and a high propensity for local recurrence after surgical excision. It arise during early to middle age and typically found on the trunk, proximal extremities, head, and neck.
Care report: A 35 year old multipara presented at gestational age of 34 weeks with epigastric mass of a year duration that progressively increased as her pregnancy advanced. Examination revealed a subcutaneous mass extending from the epigastric region with shiny, irregular, soft and tender... The symphysio-fundal height was 30 centimetres within which is a singleton foetus with audible foetal hearth rate.
She was co-managed with the plastic surgeons as a probable case of rhabdomyosarcoma and was planned for excision biopsy post delivery. She had cervical ripening with Foley's catheter at 37 weeks and 3 days but it failed and she subsequently had emergency caesarean section, bilateral tubal ligation and wide local excision. She was delivered of a live female baby that weighed 2.4 kilograms. Histology report revealed Derma to fibro sarcomaprotuberans.
Conclusion: Derma to fibro sarcoma is a rare tumour with malignant potential. It co-existed with pregnancy and was successfully managed with favourable outcome.

Keywords: cesarean section, derma to fibro sarcoma protuberance, live fetus, pregnancy, rhabdomyosarcoma

1. Introduction

Derma to fibro sarcomaprotuberans (DFSP) is a relatively unusual fibrohistiocytic tumor that is classified as an intermediate-grade malignancy [1]. It is characterised by infiltrative growth and a high propensity for local recurrence after surgical excision [1]. Despite it's locally aggressive behaviour, it has little metastatic potential [2], and when distant metastasis occur, it almost always arises after repeated local recurrences and usually involves lung, bone, or loco regional lymph nodes [1, 2].

DFSP have an incidence of 3-6.5 cases per million persons per year, and usually arise during early to middle age[2]. It is typically found on the trunk, proximal extremities, head and neck, but may present at almost any site [1, 2]. Furthermore, it have a high propensity for local recurrence. According to literature, about 11% to 53% of affected patients experience relapse after adequate wide excision, and the local recurrence rate was affected by the depth of disease invasion, site of disease, high mitotic activity, microscopically close resection margin, and increased cellularity [3]. However, metastasis only occurs in less than 6% of cases [2].The annual incidence seems to be greater in blacks than in other races[4] and it appears to affect men and women equally [4].

The tumour usually anonspecificlesion that present as a small plaque with fleshy, brownish, pinkish, or even violaceous skin discolouration that might go unnoticed and often be confused with other benign lesions. It grows slowly in this initial plaque stage with different morphological appearances [5]. It may be morphea-like, in which the lesion appears as an indurated plaque with flesh, whitish, or greyish colour. The atrophoderma-type tumour, present as a soft, depressed flesh-colored plaque with an atrophic appearance. Lastly, an angioma-type tumour, which is less common and resembles vascular lesions such as; flat angioma. As the tumour grows, it infiltrates more deeply and spread, nodules start to develop on the surface. The time taken for the transition from the plaque phase or non-protruding phase to the nodular phase is highly variable, with a range of less than 1 month to up to 50 years [5].

Uncommon pigmented variant, called Bendartumour, which is an exceedingly rare tumour that account for 1–5% of all cases of DFSP. It is characterised by a usually scanty population of dendritic melanocytes within an otherwise typical DFSP. Less than 5% of DFSPs are associated with metastases and many of them show either a fibrosarcomatous component or, much more rarely, malignant fibrous histocytoma [MFH] "like appearance" [6].

Macroscopically, DFSP appears as a single, fairly well-delimited mass in the dermis. It has a firm consistency and a yellowish or greyish colour. Macroscopically, infiltration of subcutaneous cell tissue is usually the norm. It has a well-differentiated fibrosarcoma composed of a dense and uniform proliferation of spindle cells, with large and elongated nuclei, negligible pleomorphism, and low mitotic activity[7]. The stroma has variable quantities of collagen and capillaries. One of the most important histologic characteristics of DFSP is the

arrangement of these cells in intermingled bundles in an irregular or storiform pattern [7]. In a less common presentation, the cells may be arranged radially around a central fibrous hub in a cartwheel pattern.

There are only few reports in the literature describing derma to fibro sarcomaprotuberans that enlarged considerably during pregnancy. This article describes an unusual case of the atrophic variant of DFSP with rapid enlargement during pregnancy.

2. Case Presentation

A 35 year old $G_5P^{4+0}A_5$ at34 weeks gestational age. She was referred from a peripheral hospital with complaint of epigastric mass of a year duration. The mass was initially small but progressively increased during index pregnancy to its current at about 34 weeks. There was associated pain and itching. However, there was no trauma, bleeding or discharge. On examination; she was a young woman not afebrile, not pale anicteric and acyanosed. There was no pedal oedema or peripheral lymphadenopathy. The cardiopulmonary examination was essentially normal. The abdomen was enlarged with a mass in the epigastric region, it was firm with shiny, irregular, soft and firm and surfaces. It measured 12 x 14 x 10centimetres. it was tender and the examining fingers can get above and below it. The tumour is mobile and not fixed to the underlying structures but fixed to the skin. Other organs were not enlarged. The symphisio-fundal height was 30 centimetres with a singleton fetus in longitudinal lie, cephalic presentation and the feotal heart rate was 134 beats permit minute. Pelvic examination was normal. Finding is as in image 1 below.

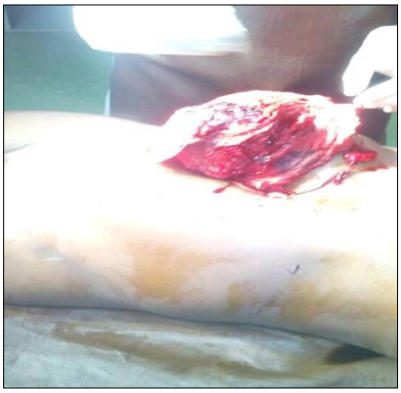

Fig 1: Anterior abdominal wall mass

She and her relation were counselled on the condition and its management, and was admitted. Investigations requested and their results were Packed Cell Volume of 28%, Clotting profile, renal function and liver function tests, RVS, VDRL, Urinalysis were essentially normal. Blood group is O rhesus D positive and haemoglobingenotype of AS. Abdomino-pelvic ultrasound scan revealed a huge thick walled cystic mass with solid components at the anterior abdominal wall. The content was turbid with septations within it there was no communication with the anterior abdominal cavity. The uterus contained a singleton live intrauterine feotus in longitudinal lie and cephalic presentation with good cardiac activity. The placenta was anterior fundal and the amniotic fluid was adequate. The estimated gestational age was 34 weeks and the. The conclusion was a complex anterior abdominal mass with solid and cystic mass coexisting with intrauterine gestation. She was continued on haematinics and monitored closely.

The plastic surgeons were invited and she was planned for excision biopsy after delivery. However, 7 days on admission the mass rapidly increased in size to about 20 x14 x 16 centimetres, it ruptured with associated bleeding. She was commenced on daily dressing, antibiotics, analgesics and steroids to enhance lung maturity. Four units of blood were crossmatched.

She was counselled on induction of labour with cesarean section if need be. Consent was otained and she had cervical ripening with Foley's catheter at 37 weeks and 3 days which failed. She subsequently had caesarean section, bilateral tubal ligation and wide local excision under general anaesthesia in conjunction with the plastic surgeons and was delivered of a live female neonate with APGAR scores of 7^1 and 9^5 and birth weight of 2.4 kilograms. She had 3 units of blood transfused intra-operatively.

The operative findings were; fungatingtumour with central necrosis at the upper part of the abdomen, the tumour is attached to the rectus sheath. The liver and abdominal organs were free of the tumour macroscopically. Operative findings are as shown in images 2 & 3 below.

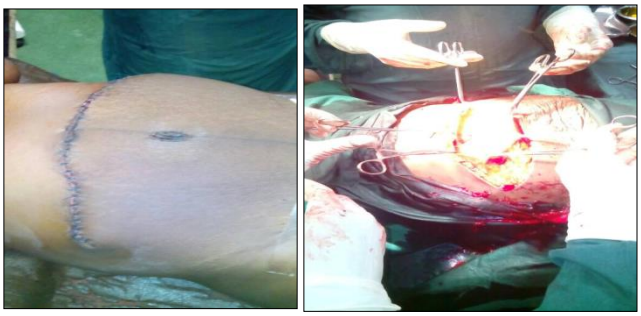

Fig 2 & 3: Post wide local excision of the lesion

Histology result revealed an ulcerated skin covered by fibrofatty tissues with accompanying grey brown fragments measuring 29 x 19 x 11 centimeters and weighed 1.23 kilograms. Cut surfaces shows a grey haemor rhagictumorextending to the deepresection margin. Microscopy revealed an ulcerated dermal tumour growing in sheets and facalst or iform patterns. The composing cells are spindly having euchromatic nuclei with eosinophilic cytoplasm. The stroma is abundant and fibrocollagenous to myxoid. The deep and lateral margins are free of the tumour. Conclusion: Derma to fibro sarcoma protuberans.

Skin biopsy showed poorly circumscribed proliferation of spindle cells that are arranged in interlacing fascicles producing storiform pattern, infiltrating the subcutaneous fat and isolating adipocytes forming honey comb or swiss cheese pattern

3. Discussion

Derma to fibro sarcoma protuberans (DFSPs) was first described by Darier and Ferrand in 1924 and named by Hoffmann in 1925. The tumour usually occur in adults of 20–50 years and rarely seen in the newborn and elderly [8].

DFSP is genetically characterised by the unbalanced chromosomal translocation t(17;22) (q21;q13), usually in the form of a supernumerary ring chromosome. The product of this

chromosomal translocation is the chimeric gene COL1A1-PDGFB (collagen type I alpha I-platelet-derived growth factor beta), which is amplified at low levels in the ring chromosome which triggers the proliferation of DFSP tumour cells through PDGF receptor tyrosine kinase [9].

The most common location is the trunk (62%), followed by the extremities (25%) and the head and neck regions (13%) [10]. The index patient presented her's on the trunk which is the most common site of occurrence.

There were only few reports in the literature describing DFSP that enlarged considerably during pregnancy. In the report, they presented 2 patients in whom derma to fibro sarcoma protuberans appeared and grew rapidly during pregnancy. Fibro sarcomatous change during pregnancy had also been described by Cakir B arising in DFSP on the scalp [11]. Three additional cases of DFSP that showed accelerated growth during pregnancy reported by Parlette [12]. The tumors in all 3 patients, and 4 additional DFSPs from 2 male and 2 female subjects, showed expression of progesterone receptor.

The index patient also showed considerable enlargement of the tumour during pregnancy. She was therefore, co-managed with the plastic surgeons during her antepartum, intra-partum and post partum period. As with many other stromal neoplasms, DFSP appear to express low levels of hormone receptors, which may be one factor that accounts for their accelerated growth during pregnancy.

The tumour is usually anonspecificlesion that present as a small plaque with fleshy, brownish, pinkish, or even violaceous skin discolouration that might go unnoticed and often be confused with other benign lesions. It grows slowly in this initial plaque stage with different morphological appearances[5]. It may be morphea-like, atrophoderma-type tumour and an angioma-type tumour. The time taken for transition from the plaque phase or non-protruding phase to the nodular phase is highly variable, and range between less than 1 month to up to 50 years [5]. The index case presented with the atrophoderma-type tumour and developed it over 8 months duration.

Metastases are rare. A review of 913 cases of DFSP described regional lymph node metastases in about 1% and distant metastases in approximately 4% [13]. The lungs are the most frequent site of metastases, but metastases to brain, bone, and heart had also been reported[12]. It usually occur within 6 years[14].

However, despite the accelerated growth in pregnancy immune histochemical studies were negative for estrogen and progesterone receptors.

Imatinib is an oral class of medications called protein-tyrosine kinase inhibitors that inhibits the platelet-derived growth factor receptor (PDGF-R) signalling cascade which plays a crucial role in the pathogenesis and tumour growth of DFSP. Imatinibmesylate is indicated for the treatment of adult patients with unresectable, recurrent, and or metastatic DFSP. A response rate of approximately 65% has been achieved among DFSP patients treated [15]. However, such is not readily available in our environment for such purpose.

Mohs micrographic surgery (MMS) with a surgical margin of 2.5 cm deep to fascia may be the treatment of choice for DFSP, because of its high cure rate and maximal conservation of tissue. The case presented had wide local excision of the tumour with satisfactory tumour margin.

Most local recurrences, which can occur in 20–49% of cases, are noted within the first 3 years after excision, but late recurrence had been reported as well [10]. The recurrence rate is the highest in DFSP of the head and neck because of cosmetic and functional restrictions of resecting large areas [10]. The greater the number of recurrences, the more likely the risk to disseminate [10]. As reported in many studies, the recurrence rate with MM was 0.6–1.6%, while the average recurrence rate was 18% when treated with wide excision [11].

The atrophic variant of DFSP could be misdiagnosed as morphea, therefore dermatologist should be aware of this uncommon but characteristic presentation and a close follow up of the lesions during pregnancy for sarcomatous changes.

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