Synovial Sarcoma In The Anterior Abdominal Wall Of A Young Nigerian Female: Case Report.

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Citation

Abstract
Synovial sarcoma is a malignant mesenchymal neoplasm so named after its microscopic resemblance to normal synovium. Large joints of the extremities are the most common site and, rarely, the tumor may be found in the anterior abdominal wall. We report a case of synovial sarcoma in the right upper quadrant of the anterior abdominal wall of a 21-year-old Nigerian female. The patient underwent surgical excision with clear margins and the histological and immunohistochemical studies confirmed a monophasic synovial sarcoma.

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INTRODUCTION
Synovial sarcoma is a malignant mesenchymal neoplasm with dual epithelial and mesenchymal spindle cell morphology. It commonly occurs in the individuals in their second to fifth decades of life. The first two cases of the tumor arising in the abdominal wall were reported by Pack and Ariel in 1950. A literature search showed that up to 2005, a total of 44 cases were reported in English literature with about 30 cases reported between 1978 and 2005. We report the first case of histologically confirmed anterior abdominal wall sarcoma in a Nigerian patient.

CASE REPORT
A 21-year-old previously healthy Nigerian female presented for medical care in a mission hospital in Onicha-Ngwa, a suburban part of Abia State, Nigeria. She noticed a non-tender, swelling in the right upper quadrant of her abdomen two years prior to presentation with gradual increase in size. She reported no gastrointestinal symptoms or weight loss. The mass was located within the muscular layer of the right upper quadrant of the anterior abdominal wall and measured 15 by 6 centimeters. It was ovoid in shape and firm but with a soft center. The overlying skin was mobile. No additional masses were found on physical examination and imaging. She was healthy looking and had no jaundice or signs of anemia.

Preoperative investigations were limited to complete blood count (within normal limits), urinalysis (within normal limits) and non-fasting blood glucose (87 milligrams/deciliter). An abdominal ultrasound revealed a mixed echogenic mass arising from the anterior abdominal wall.

Intra-operatively, the encapsulated mass was firm and appeared to arise from the oblique muscles or, possibly, the rectus muscle. There was hypervascularity of the parietal peritoneum but the tumor did not grossly penetrate into the abdomen. The aponeuroses and muscle groups bordering the mass appeared grossly normal. The tumor was completely excised with a 2 cm surgical margin, which included portions of the rectus sheath and the parietal peritoneum. The overlying subcutaneous tissue and skin were spared and used for wound closure. The defect was reconstructed with polypropylene mesh which was covered with the overlying skin and subcutaneous tissue (figs. 2). The bisected tumor showed multi-cystic blood-filled areas and some pale solid portions (fig. 3). The histo-pathological report demonstrated a monophasic (fibroblast-like spindle cell) synovial sarcoma (figs. 4 and 5) with a negative margin. Immunohistochemical
stain showed a strong positive reaction to monoclonal antibody to TLE1 (fig. 6). The tumor was also positive to EMA and CD34 (fig. 6). Following surgery, the patient had an uneventful course. At three-month follow-up there is no sign of recurrence, the wound is well-healed, and she is preparing for radiation therapy (fig. 7).

Figure 1
Figure 1: Right upper quadrant abdominal mass in a 21-year-old female pre-operatively showing a protuberant fixed mass (C = Chest, L = left, R = Right, U = Umbilicus).

Figure 2
Figure 2: Right upper quadrant abdominal wall defect reconstructed with polypropylene mesh for closure after excision of mass and musculature.

Figure 3
Figure 3: Bisected surgical specimen showing areas of fibrous white tumor admixed with hemorrhage.

Figure 4
Figure 4: Photomicrograph of monophasic fibroblast-like spindle cell areas of the tumor; no epithelioid components were identified (Hematoxylin & Eosin stain, 400x)
DISCUSSION

Synovial sarcoma is a malignant mesenchymal tumor of dual epithelial and fibroblast-like spindle cell differentiation potentials. It is known to occur most commonly in extremities, particularly in the lower ones, in close association with the joints, but not involving them \cite{1,2,6}. Nevertheless, it occurs in other parts of the body, and Adebayo and his co-workers reported a case in the nasopharynx in Northern Nigeria in 2005 \cite{7}. It rarely occurs in the anterior abdominal wall \cite{1,2,8} but, when it does, it usually affects the lower quadrants \cite{3,9}. It is usually a slow-growing, painless mass with no associated gastrointestinal symptoms.

Diagnosis based on clinical grounds is almost impossible and complete surgical excision with histological examination is required. There can be intralesional haemorrhage (fig.3) which can suggest a cystic mass on physical examination. Radiological investigations may demonstrate a heterogenous septate mass with mixed solid and cystic appearance; calcifications may be seen in about a third of cases. Magnetic resonance imaging (MRI) delineates the mass from the surrounding normal tissue and can show neurovascular or regional lymph node involvement \cite{10}. In Nigeria, as in many parts of Africa with resource-confined setting, many of these diagnostic and confirmatory tests, including immunohistochemical and molecular analyses, are either not affordable by patients or not available to physicians. Therefore, a high index of suspicion is of utmost importance when planning for surgical interventions, in order to avoid recurrence and progression of disease.
Overall 5-year survival of synovial sarcoma is reported to be between 50 and 80%, depending on the age of the patient, tumor size (better for size <5cm in diameter) and resectability[4]. National Comprehensive Cancer Network (NCCN) guidelines recommend total surgical resection with at least a 1 centimeter margin or intact fascial margins, with preoperative or postoperative radiation and chemotherapy using Ifosfamide-based chemotherapy[11]. Prior to NCCN guidelines in 2007, there was no established minimal margin of resection[12]; however, many investigators have suggested a tumor-free margin of 1-3 centimeters in adults[12,13,14]. Asher et al. report a three-year follow-up without recurrence or metastasis after surgical resection of a synovial sarcoma of the vulva in a 28-year-old Caucasian lady, without adjuvant therapy[10]. Similarly, Ozaydin and Aslaner[8] reported a three-year follow-up without evidence of recurrence or metastasis after surgical resection of an anterior abdominal wall synovial sarcoma, followed by adjuvant radiotherapy (50Gy).

Bearing in mind that inadequate surgical margins, among other prognostic factors, contribute to bad outcomes in synovial sarcoma, we believe that wide surgical resection should be the focus in resource-confined settings in order to achieve the best outcomes.

CONCLUSION

Synovial sarcoma of the anterior abdominal wall is rare. This first case report of a patient from Nigeria demonstrates wide surgical margins of excision which should be the standard in resource-confined settings. We would predict an excellent prognosis in our patient, given her age and surgical-margin status.

References

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