EDITORIAL

Congenital Lower Limb Biphasic Synovial Sarcoma in a Male newborn - Khartoum Teaching Hospital: A Case Report

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Abstract:

Synovial sarcoma is a malignant mesenchymal neoplasm which commonly occurs in the extremities of adults. We report on a 12-days-old boy who presented to the department of paediatric surgery at Khartoum teaching hospital with a biphasic synovial sarcoma in the lateral aspect of his left ankle. The tumor was treated with total excision and adjuvant chemotherapy.Histopathologic examination of the excised tumor revealed a biphasic synovial sarcoma. There was no evidence of local recurrence or distant metastasis of the tumor at follow-up examination 24 months postoperatively.

Key words: Biphasic synovial sarcoma, congenital, lower limb

Introduction:

Soft-tissue sarcomas are a relatively rare disease accounting for approximately 1% of adult malignancies [(1)]. Synovial sarcoma, one of the most common types of soft tissue sarcomas, usually presents in the proximal or middle portions of the extremities, often as a large mass with an aggressive clinical behavior. Although synovial sarcoma can occur in any part of the body, more than 80% of tumors arise in the deep soft tissue of the extremities, especially around the knee. The etiology of synovial sarcoma is unknown but they appear to have an epithelial phenotype [(2)]. Although it is known for being particularly aggressive, synovial sarcoma often grows slowly, forming a circumscribed, multinodular tumor without a capsule [(3)]. Gland-forming biphasic and spindle cell fibrous monophasic tumors are the most common subtypes. [(4)] Tumor size, depth, invasion, and primary location affect survival in pediatric cases. [(5)]
Surgical excision with wide, negative margins is the currently recommended treatment with adjuvant radiotherapy and/or doxorubicin-based chemotherapy [(6)]. While this is a mainstay of treatment, there is no consensus on the optimal treatment strategy

Case report:

A 12 days old boy presented with swelling of the left foot since birth which increased in size gradually with yellowish discharge. No history of neoplasia in the family. His weight was 3 kg and he was not pale.
Local examination showed a cystic mass at the lateral aspect of the dorsum of the left foot over laying the ankle joint, 3 by 6 cm and its center was covered with blackish crust. (Figure 1: a & b)
The Hb was 13 gm/dl, Urea 20 mg/dl, creatinine 1.2 mg/dl, sodium 131 mmol/L and potassium 3.8 mmol/L.
X-Rays showed normal bones of both feet

EDITORIAL

The mass was excised with healthy margins and the defect closed primarily after undermining of the edges. The histopathology report showed a tumor composed of polygonal & spindle shaped cells. The polygonal cells have large nuclei with punctuate chromatin. In some foci they form glands. The spindle shaped cells have dark nuclei & arranged in edematous stroma .The tumor is infiltrating the skin where there is ulceration. There are scattered mitoses. The glands are positive for the Epithelial Membrane Antigen (EMA). All the cells are positive for Vimentin and negative for Desmin. Features are typical of a biphasic synovioma.

He went through smooth post operative period and received radiotherapy as adjuvant treatment. Follow up for 2 years since the operation showed no local recurrence or distal metastasis.

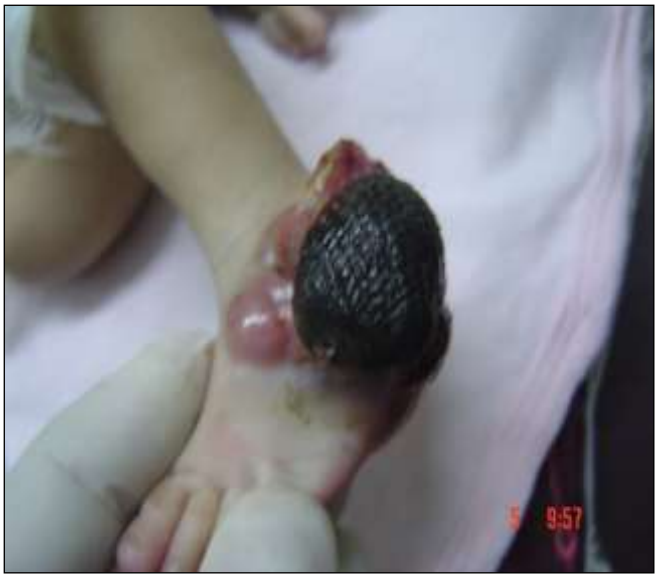

Figure 1- a: Local examination of the tumour

EDITORIAL

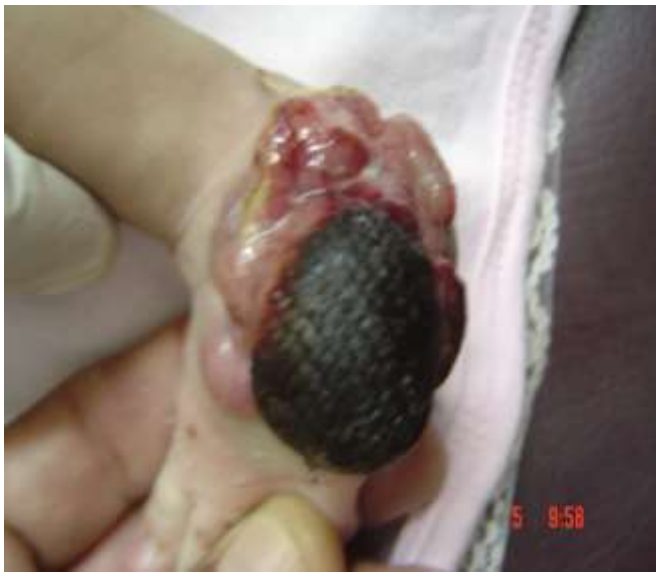

Figure 1 - b: Local examination of the tumour

Discussion:

Synovial sarcoma is a malignant neoplasm of soft tissue that typically arises near large joints of the upper and lower extremities of young adult males, particularly the knee; however, they do not arise from synovial tissue [7,8] but from malignant degeneration of primitive mesenchymal cells [8]. The microscopic appearance of the degenerated mesenchymal cells is remarkably similar to synovial tissue, hence the name of the tumor. [9]

Synovial sarcomas are usually treated aggressively with wide excision with negative margins, often including removal of adjacent muscle groups and even total amputation [7]. Limited excision is unfortunately associated with a high incidence of local recurrence (60-90%) within 2 years of the original surgery [10].In another report, synovial sarcomas were the most common extremity lesions, and one-third of the patients develop locally recurrent disease with a median disease-free interval of 18 months [1]. The surgical excision is followed by post-operative radiotherapy and chemotherapy to help control metastasis. [11]

Complete surgical excision with safe margins and adjuvant radiotherapy was the option of treatment in our case and the outcome is free local area up to the 24 months follow up visit, and this can be fairly compared to the results of Yokoyama H who reported a case of agirl of 13 months with biphasic synovial sarcoma. [12]

Conclusion:

Best available evidence suggests that a multimodal treatment strategy starting with aggressive surgical resection followed by radiation and chemotherapy offers the patients the best chance for a cure.

EDITORIAL

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