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Parveen Kumar*

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Case Report

The Diagnostic Pitfall for Angiosarcoma in one year old child with Extremity Tumor

Parveen Kumar[1*], Lavleen Singh[2], Geetika Mathur[1]
[1]Department of Pediatric Surgery, Chacha Nehru Bal Chikitsalya, New Delhi, India.
[2]Department of Pathology, Chacha Nehru Bal Chikitsalya, New Delhi, India.
***Corresponding Author:** Parveen Kumar, Department of Pediatric Surgery, Chacha Nehru Bal Chikitsalya, New Delhi, India.
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Abstract

Pediatric angiosarcomas are very rare and less studied. There exists a significant degree of confusion in histologically differentiating angiosarcomas from other endothelium derived tumors. We present here a case of right forearm mass in an infant with diagnostic dilemma, which later turned out to be angiosarcoma.

Keywords: pediatric; infant; rhabdomyosarcoma; endothelium; vascular tumor

Introduction

Angiosarcoma is an aggressive malignant endothelial cell tumor of lymphatic or vascular origin. They are rare tumors that usually affects adults/elderly patient. Pediatric angiosarcomas are exceedingly rare, poorly studied and a very little is known about its line of treatment. The classic presentation is an enlarging, painful mass of several weeks' duration and usually associated with anemia and/or thrombocytopenia.[1] Due to the variability in the appearance of angiosarcoma, the correct diagnosis can often be delayed. It is characterized by polymorphic and non-specific clinical and radiological features. These lesions may have a destructive pattern on radiology. Immunohistochemistry support is often needed as vasoformative architecture usually get missed on light microscopy. Treatment modalities are variable depending on tumor extent, resectability and metastasis.[1] The prognosis is often dismal.

Case details

A 1-year-old male child presented with right forearm mass for 6 months. It was progressively increasing in size associated with foul smelling watery discharge [Fig.1a]. The incisional biopsy had more of necrotic tumor, causing interpretation of IHC difficult. The tissue had spindle cells and desmin positivity and was reported as suspected case of sarcoma possibly embryonal RMS. Contrast enhanced computer tomography (CECT) showed soft tissue density mass lesion in muscular layer of lateral and posterior compartment of right forearm, with encasement of neurovascular bundle and elbow joint, multiple enlarged supra-trochlear lymph nodes. FDG PET CT showed heterogenous enhancing mass with involvement of skin. FDG uptake was positive in supra-trochlear lymph nodes, right axillary lymph nodes, occipital and sphenoid bones.

Figure 1a: Right forearm mass

He received 12 weeks of VAC (Vincristine, Actinomycin D and Cyclophosphamide) neo-adjuvant chemotherapy and had partial response (>50% size reduction) [Fig.1b]. He underwent above elbow amputation for mass and histo-pathological examination showed tumor with ramifying vascular spaces, showing mild to moderate atypia [Fig. 2a, 2b]. The vessels were diffusely infiltrating into dermis, extending into subcutaneous tissue and skeletal muscle. The tumor cells were immune-positive for CD31, CD34 and D2-40 and final reporting was angiosarcoma [Fig.2c]. Post-operatively he developed edema of amputated stump and later child succumbed to sepsis secondary to neutropenia.

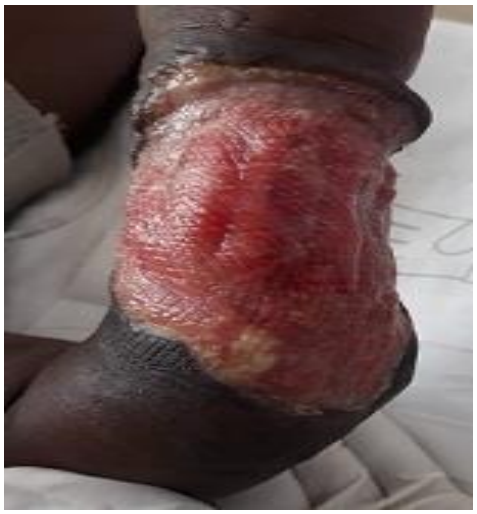

Figure 1b: Partial response status post neo-adjuvant chemotherapy

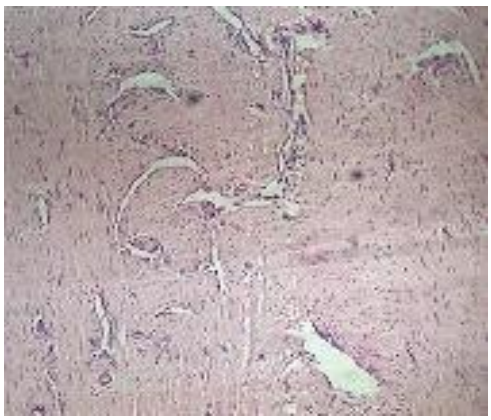

Figure 2a: Photomicrograph showing tumor comprising of ramifying vascular spaces which are lined by hobnail endothelial cell (H&E;100)

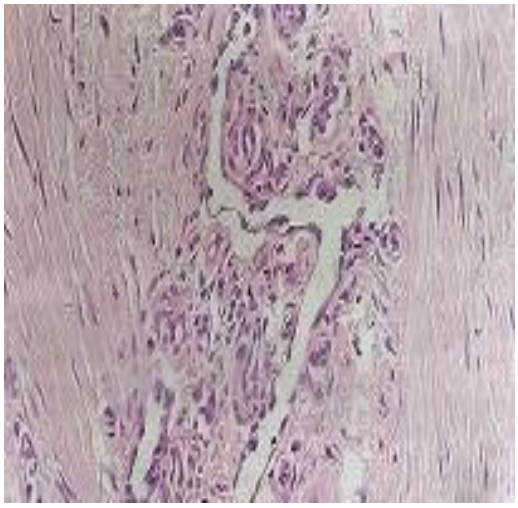

Figure 2b: Endothelial cell lining the vascular spaces shows mild to moderate atypia.

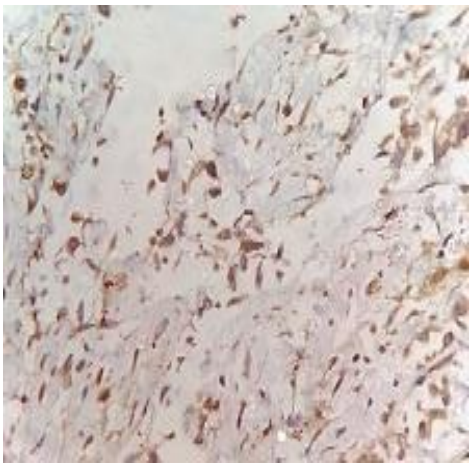

Figure 2c: Endothelial cell show positivity for CD 34

Discussion

Endothelium derived neoplasms range from benign hemangiomas to tumours of intermediate malignancy such as hemangioendothelioma to highly malignant and aggressive angiosarcomas (AS).[2] There exists a significant degree of confusion in histologically differentiating these tumors. AS may arise spontaneously.[2] It has been reported in areas of chronic lymphedema, areas of local radiation and exposure to foreign materials with chronic inflammation. So, it is difficult to establish risk factors.

AS account for 1% and 2% of all sarcomas; around half are cutaneous and elderly predominance.[3,4] Most common sites being head and neck. It is extremely rare in childhood. Age at diagnosis range from 3 months to 16 years.[5] Youngest age for AS in literature is in 17 days old neonate.[6] Pediatric AS has slight male predilection.[7] The two most common etiological factors are chronic lymphedema and radiation therapy.[8] Histopathology shows various extents of differentiation with atypia of endothelial cells and vague vessel formation. Hobnail morphology is rarely seen.[9] The immunohistochemical panel for AS includes a basic panel for spindle-cell tumors (CD31, pancytokeratins, S110, and actin) and additional vascular markers (CD34, erythroblast transformation-specific-related gene [ERG], podoplanin).[10] In our case, the initial incisional biopsy had more of necrotic tumor, causing interpretation of IHC difficult. The tissue had spindle cells and desmin positivity and was reported as suspected case of sarcoma possibly embryonal RMS. So, patient received VAC chemotherapy followed by above elbow amputation. The histopathological examination showed presence of vague vessel formation, giving an impression of vaso- formative tumor on histology. IHC was supportive with positivity of CD31, CD34 and D2 40 and final reporting was angiosarcoma. AS should be kept in mind while dealing with vascular tumors having pleomorphism, mitotic activity and necrosis. The vaso-formative nature may be missed on microscopy, so need supportive IHC for diagnosis.

The complete surgical excision is the mainstay of treatment in localised tumor, if feasible. Unfortunately, upto 45 % of AS are metastatic at presentation.[11] Study by Ferrari et al demonstrated that 25% of pediatric AS were metastatic at diagnosis.[7] AS spreads primarily by hematogenous route to lungs, liver, bone and lymph nodes.[12] Due to metastatic presentation and various limitations for surgery, various adjuvant therapeutic modalities may be employed. Adjuvant radiotherapy has been demonstrated to prolong survival.[11] It can be used alone or in combination with chemotherapy. Adjuvant chemotherapy may provide initial clinical responses but final outcome remains poor. Our case did not receive radiotherapy and succumbed to chemotherapy even after surgical

excision. Radiation therapy may play a role, but the greater risks posed to children by treatment should be considered.

Fayette et al studied 161 AS patients treated from 1980 to 2004 and found that primary sites were breast (35%), skin (20%) and soft tissues (13%) with a median age of 52 years. 31 cases (19%) had metastases at initial presentation. The most common metastatic sites at initial diagnosis were lungs (25%), bone (22%) and liver (16%). R0 resection was possible in 85 of 121 operated patients (localised disease n=128).[3] In spite of localised disease, 11.1% had microscopically positive margins out of 821 operated cases from 2004 to 2012.[4] Increasing age, black race, grade 3 histology, increasing size and margin status were found to be significantly associated with poor overall survival.[4] It was consistent with other studies in literature.[13-16] Literature also quotes poor prognostic factors including size > 5 cm, positive resected margin, metastasis and presence of necrosis [3]. It has 5 year survival rates ranging from 12% to 15%.[8] The surgery in AS may be problematic due to multicentre/multifocal character of disease or poor clinical delimitation and are often diagnosed late or with metastasis.[10] Even with localised disease, only 60% of AS patients survive for more than 5 years, with a median survival of 7 months.[17]

Conclusion

Pediatric angiosarcoma are more aggressive as compared to adult counterparts. Pathological diagnosis is difficult and require high expertise with immunohistochemical study to confirm vascular differentiation. The role of chemotherapy is uncertain, with complete surgical excision as mainstay of treatment, albeit possible in only few cases. Post-operative radiotherapy may have a role in local control.

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