A red nodule on the tip of the nose in a Chinese girl

A 7-year-old girl presented with a red nodule on the apex of the nose for 1 month without any other symptom. Skin examination revealed a well-defined, round, pink nodule measuring 1.2 × 1.5 cm with a hard texture [Figure 1a]. Her past medical history was unremarkable.

An incisional biopsy showed normal epidermis and nests or sheets of small round cells with atypical nuclei and scanty cytoplasm in the reticular dermis. The tumor cells formed alveolar structures. Necrosis, cross striations or obvious mitotic activity were absent [Figures 1b and c]. Immunohistochemical studies demonstrated that the tumor cells were strongly positive for vimentin, myoblast determination protein 1, desmin and myogenin, weakly positive for myoglobin (focal), synaptophysin, cytokeratin and Ki-67 (30% +); and negative for S-100, smooth muscle actin and cluster of differentiation (CD) 3 [Figures 2a-d].

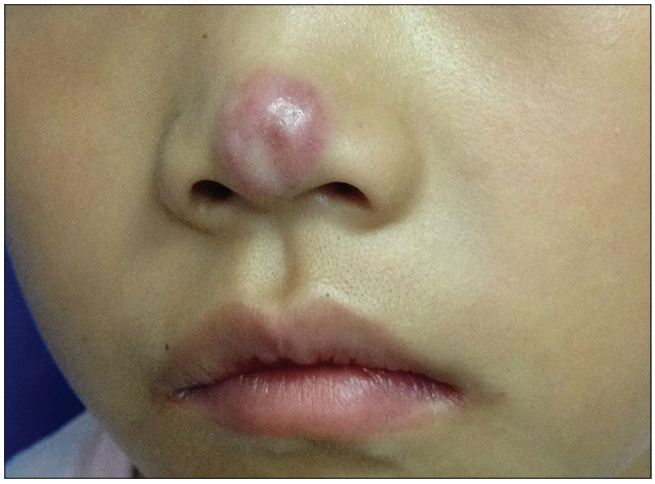

Figure 1a: Solitary, pink nodule on tip of nose of a 7-year-old girl

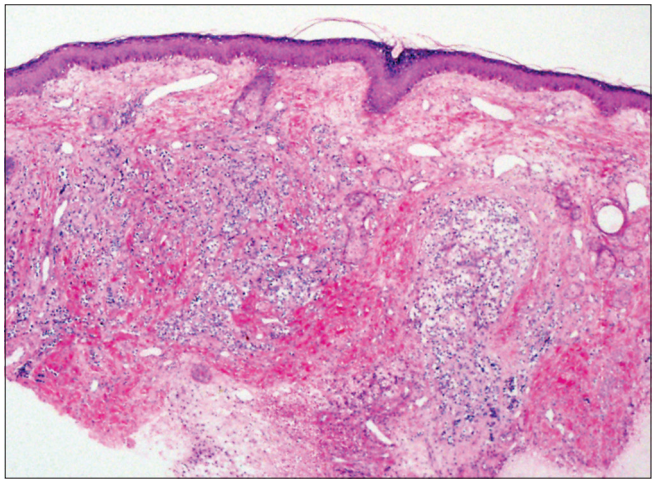

Figure 1b: Nest or sheet-like distribution of tumor cells with atypical nuclei and scanty cytoplasm, with some tumor cells forming alveolar structures. No necrosis, cross striations or obvious mitotic activity was found (hematoxylin and eosin, × 40).

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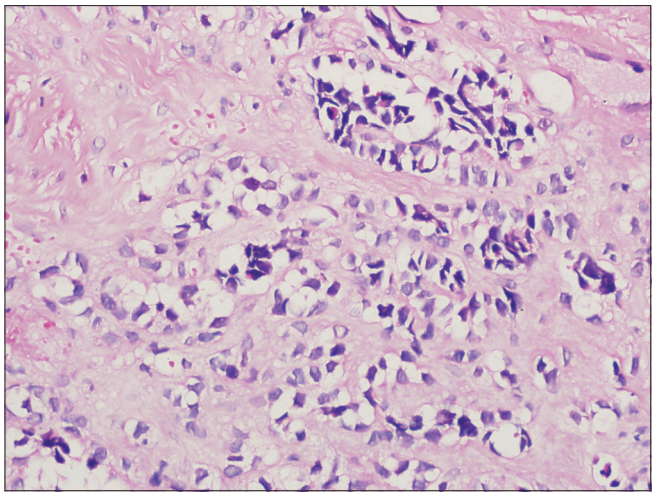

Figure 1c: Nest or sheet-like distribution of tumor cells with atypical nuclei and scanty cytoplasm, some tumor cells forming alveolar structures. No necrosis, cross striations or obvious mitotic activity was found (hematoxylin and eosin, × 400).

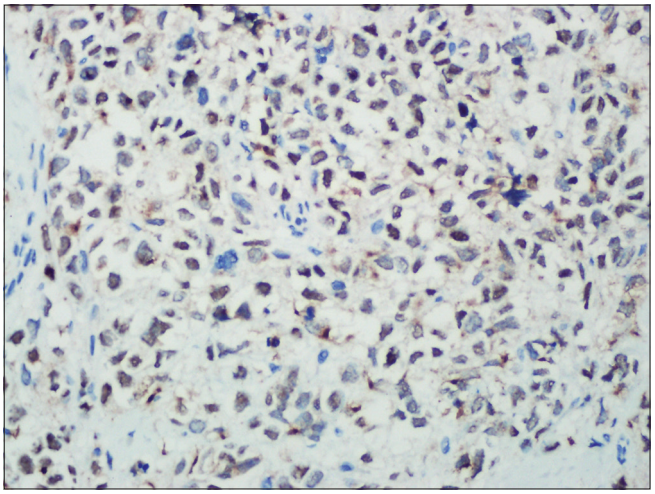

Figure 2a: Immunohistochemical stain showed reactivity for myoblast determination protein 1 (×400)

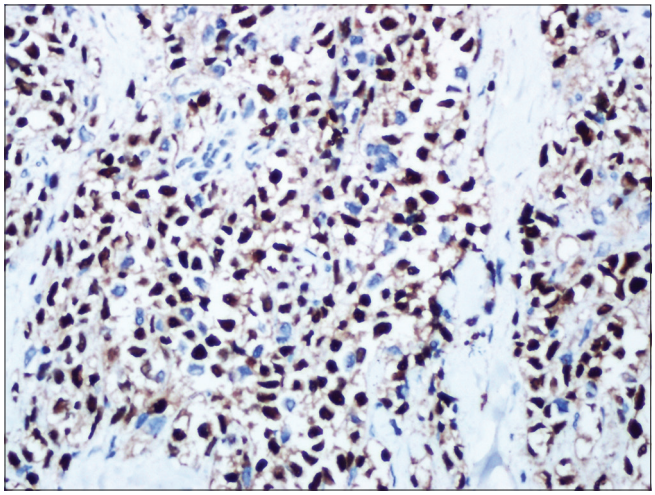

Figure 2b: Immunohistochemical stain showed reactivity for myogenin (×400)

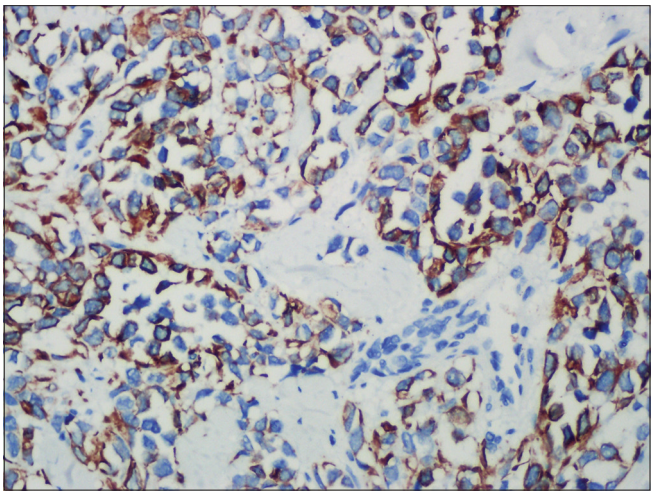

Figure 2c: Immunohistochemical stain showed reactivity for desmin (×400)

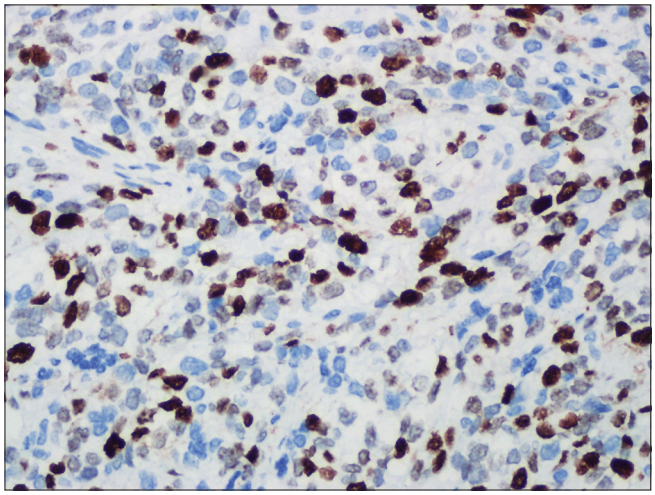

Figure 2d: Immunohistochemical stain showed reactivity for Ki-67 (30% +) (×400)

Answer

Alveolar rhabdomyosarcoma

Discussion

Rhabdomyosarcoma is the most common soft tissue sarcoma among children.[1] Rhabdomyosarcoma is classified into four types based on histological features, namely, alveolar rhabdomyosarcoma, embryonal rhabdomyosarcoma, spindle cell rhabdomyosarcoma and pleomorphic rhabdomyosarcoma. The most common site of occurence of rhabdomyosarcoma is the head and neck region. Almost all rhabdomyosarcomas occur in the deep soft tissue, on the trunk and extremities.[2] It rarely occurs on the skin and superficial dermis, as primary sites.[3]

Rhabdomyosarcoma in the head and neck region can be divided into three subgroups: orbital, parameningeal and nonorbital nonparameningeal.[2] Almost all nonorbital nonparameningeal rhabdomyosarcomas are reported to occur in deeper tissue. Rhabdomyosarcoma in the nose usually appears in the nasal cavity and rarely occurs on the skin as the primary site (comprising less than 1% of all rhabdomyosarcoma).[3] Based on histologic features, alveolar rhabdomyosarcoma and embryonal rhabdomyosarcoma are common subtypes. Usually, alveolar rhabdomyosarcoma shows more aggressive clinical behavior with a worse prognosis than embryonal rhabdomyosarcomas.[3] Till now, very few cases of superficial primary cutaneous alveolar rhabdomyosarcoma have been reported on the nasal skin.[4]

Histopathologically, the present case exhibited nests and sheets of small round tumor cells in the superficial dermis. Focal alveoli-like structures were observed, which is an important histological feature of primary cutaneous rhabdomyosarcoma.[3] These morphological features mimic melanocytic as well as epithelial tumors. The tumor cells were strongly positive for vimentin, myoblast determination protein 1, desmin, myogenin and negative for S-100, smooth muscle actin, leukocyte common antigen and CD3. The strong expression of myogenin and myoblast determination protein 1, the two most reliable markers for rhabdomyosarcoma, helped us reach a diagnosis of primary cutaneous superficial alveolar rhabdomyosarcoma and excluded malignant melanoma, Spitz nevus, vascular tumors, squamous cell carcinoma, lymphoma, leukemia, Merkel cell carcinoma, leiomyosarcoma and hematological malignancies.

The optimal treatment of rhabdomyosarcoma includes wide local excision with lymph node dissection, along with postoperative radiation and chemotherapy.[5] However, the optimal width and impact of the margin of resection have not been well-defined. The following factors have contributed to patients' outcomes, such as tumor size, histological subtype, invasion to the adjacent tissues, the involvement of regional lymph nodes and distant metastases. Due to financial difficulties, the patient did not undergo postoperative radio-chemotherapy and died of cervical metastasis a year later.

In summary, this case had an unusual presentation that could be easily misdiagnosed as Spitz nevus or another cutaneous neoplasm. The correct diagnosis of alveolar rhabdomyosarcoma was reached based on the histopathological features and immunochemistry findings. The patient's general condition rapidly worsened due to regional metastasis and died a year after the initial presentation, underlying the importance of awareness of such a disease presentation and prompt diagnosis. Optimal treatment is yet to be defined and should be based on clinical analysis of more such cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that child's names and initials will not be published and due efforts will be made to conceal the identity but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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