Recurrent periorbital low-grade myofibroblastic sarcoma in an infant

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Low-grade myofibroblastic sarcoma (LMFS) is a rare, intermediate grade tumor of myofibroblast cell origin, mostly found in adults. According to a major series, average age of presentation was 40 years (range 9–75 years); 38.7% (24 of 65) occurred in the head and neck region which included lower jaw, sinus, and oral cavity, whereas 41 cases were found in the limbs and other bones, etc.[1]

We hereby report a 10-month-old girl with a reddish lesion, painless progressive, started at the age of 1 month near medial canthus. The baby was born at full-term with uneventful antenatal and postnatal history. No history of similar lesions elsewhere in the body. A 3-month course of oral propranolol was given in the past with a provisional diagnosis of capillary hemangioma with no response. On examination, a solid, firm, non-compressible mass with involvement of skin, reddish in color, with telangiectatic blood vessels was seen, with fixity to underlying bone. It measured 30 mm × 40 mm, located in medial periorbital area [Fig. 1a]. Ocular examination was normal. MRI with contrast showed an intensely enhancing mass with hyperintense T1-weighted and isointense T2-weighted signals [Fig. 1b]. Biopsy was suggestive of LMFS. On light microscopy, fascicles of spindle-shaped tumor cells, with focal storiform pattern, were seen infiltrating soft tissues. Tumor cells had ill-defined eosinophilic cytoplasm and fusiform nuclei. Mild nuclear atypia was seen along with mitotic figs. Ki67-labeling index was >10%. On immunohistochemical

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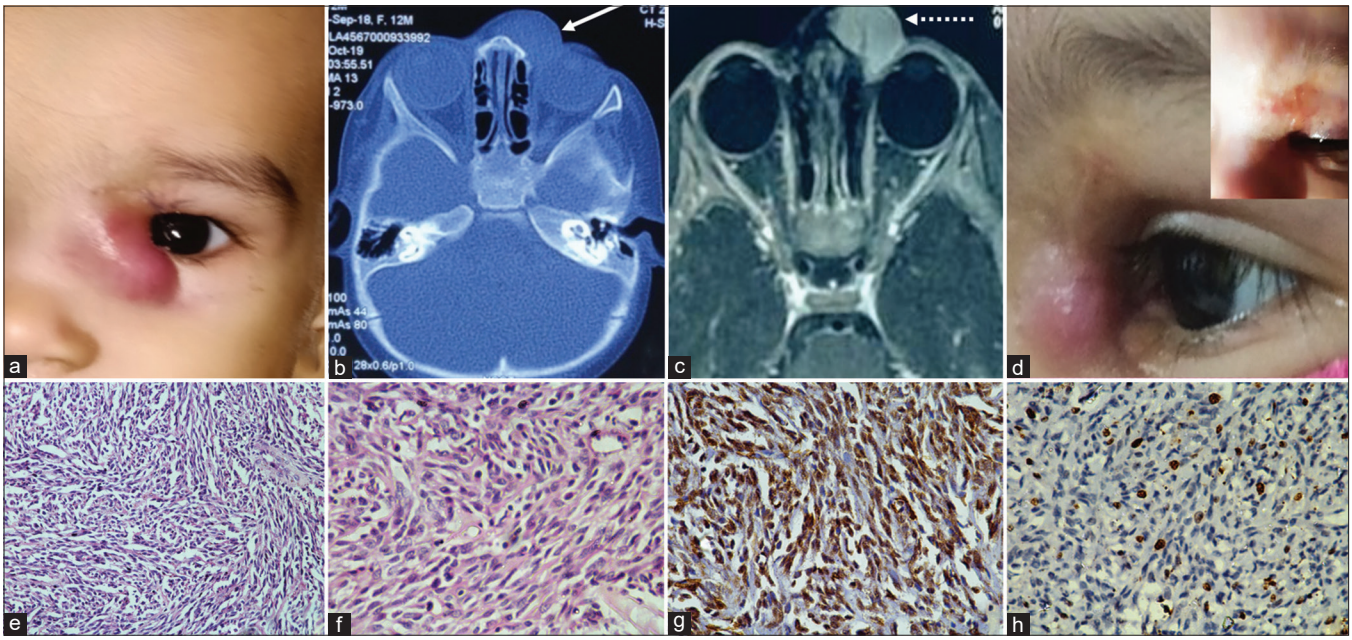

Figure 1: (a) Patient with reddish colored lesion in the left periorbital area, medially, at presentation; (b)MRI (contrast-enhanced, fat suppressed T1-weighted) shows intensely enhancing lesion with no defined borders (white dotted arrow); (c)Patient with recurrence of tumor at the same site at 6 months, follow-up; (d)Patient on oral pazopanib for 7 months duration, after repeat surgery with no recurrence; (e)Low power view shows spindle shaped tumor cells arranged in fascicles (H & E stain, ×200); (f)Higher magnification shows tumor cells have ill-defined pale cytoplasm and fusiform nuclei with mild pleomorphism. Occasional mitotic figures are seen (H & E stain, ×400); (g)Immunohistochemistry shows cytoplasmic positivity for smooth muscle actin (×400); (h)Immunohistochemistry shows Ki67-labeling as >10% (×400)

stains, tumor cells showed cytoplasmic positivity for smooth muscle actin and vimentin [Fig. 1e-h]; negative for desmin, myogenin, and S100.

Medical oncologist's opinion was also sought for neo-adjuvant chemotherapy but LMFSs are mostly resistant to chemotherapy. We planned excision of tumor along with the involved skin. Intra-operatively, tumor was found to be non-encapsulated, solid, and highly vascular. Gross total resection along with the involved skin was performed. Histopathology examination was consistent with LMFS. After 6 months of surgery, patient presented with recurrence at same site [Fig. 1c]. Although the recurrence in skin appeared small, the surrounding area was firm on palpation suggestive of infiltration by the tumor. A repeat MRI was done. Re-surgery was performed, the involved skin was excised, and dissection was carried out in subcutaneous plane with piecemeal removal of tumor done till healthy tissue was visible; superiorly till medial end of eyebrow; medially till contralateral side of nasal bridge; and laterally till medial one third of upper and lower eyelids. Local application of mitomycin-C 0.04% for 5 min soaked in a cotton piece over exposed bone and subcutaneous area was done. A glabellar rotational forehead flap was raised to reconstruct defect. Histopathology showed similar findings. In view of residual tumor, patient was started on tablet pazopanib, 100 mg Once a day (OD), by the medical oncologist. Patient is on the treatment for 7 months with no recurrence [Fig. 1d].

Discussion

LMFS are rare in pediatric age group and even rarer in first decade of life; approximately, 10 cases of LMFS have been reported in the first decade of life but none in infancy (present case at the age of 1 month).[2] Although 2 case reports of LMFS are available in ophthalmic literature, 1 in orbit and another at limbus, respectively, none avaialble in the periorbital region.[3,4]

We hypothesize that the tissue of origin in our case was cutaneous with subcutaneous infiltrative spread. Because of rarity of LGFS and wide variety of tissue of origin, that is, soft tissues, bone, or organs, imaging characteristics of LGMS is less described. In the present case, though the lesion appeared mostly well-defined, intraoperatively, the tumor was not encapsulated with ill-defined edges. Underlying bone was normal. LGMF sarcomas arising from bone show osteolytic and rarely osteoblastic changes along with the mass lesion.[2,3]

Primary treatment of LMFS is a wide-surgical excision.[1,3,4] Midline of face has important anatomical structures in close proximity, that makes wide-excision of tumor difficult, as was in the present case. Periorbital area, especially near the medial canthus, is considered high risk location for recurrence and hematogenous spread of tumor because of reasons such as, close proximity of tumor to underlying bone, high vascularity, and difficult to perform wide excision due to important anatomical structures in close proximity. Local recurrence reported in LGMF sarcoma is 29.1% (23/79), usually within 6 months to 7 years. Hematogenous spread is reported in 17.7% (14/79); lungs and bones being common sites.[1] Chemotherapy in the form of ifosfamide, vincristine, and etoposide has been attempted in the early cases but with no response.[5] However, recent newer drugs, pirarubicin and nidaplatin along with ifosfamide, was used as adjunctive treatment after surgical excision in pancreatic LGMS which showed no recurrences till 5 years of follow-up.[6] Radiotherapy

in doses of 45d Gy-66Gy was given for residual tumor which resulted in good response.[7] Pazopanib is an antiangiogenic molecularly targeted drug with effective role in different types of sarcomas.[8]

To the best of our knowledge, this is the first report of occurrence of LMFS that started at the age of 1 month in periorbital region. Further, role of pazopanib to prevent recurrence needs to be emphasized in a recurrent LMFS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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