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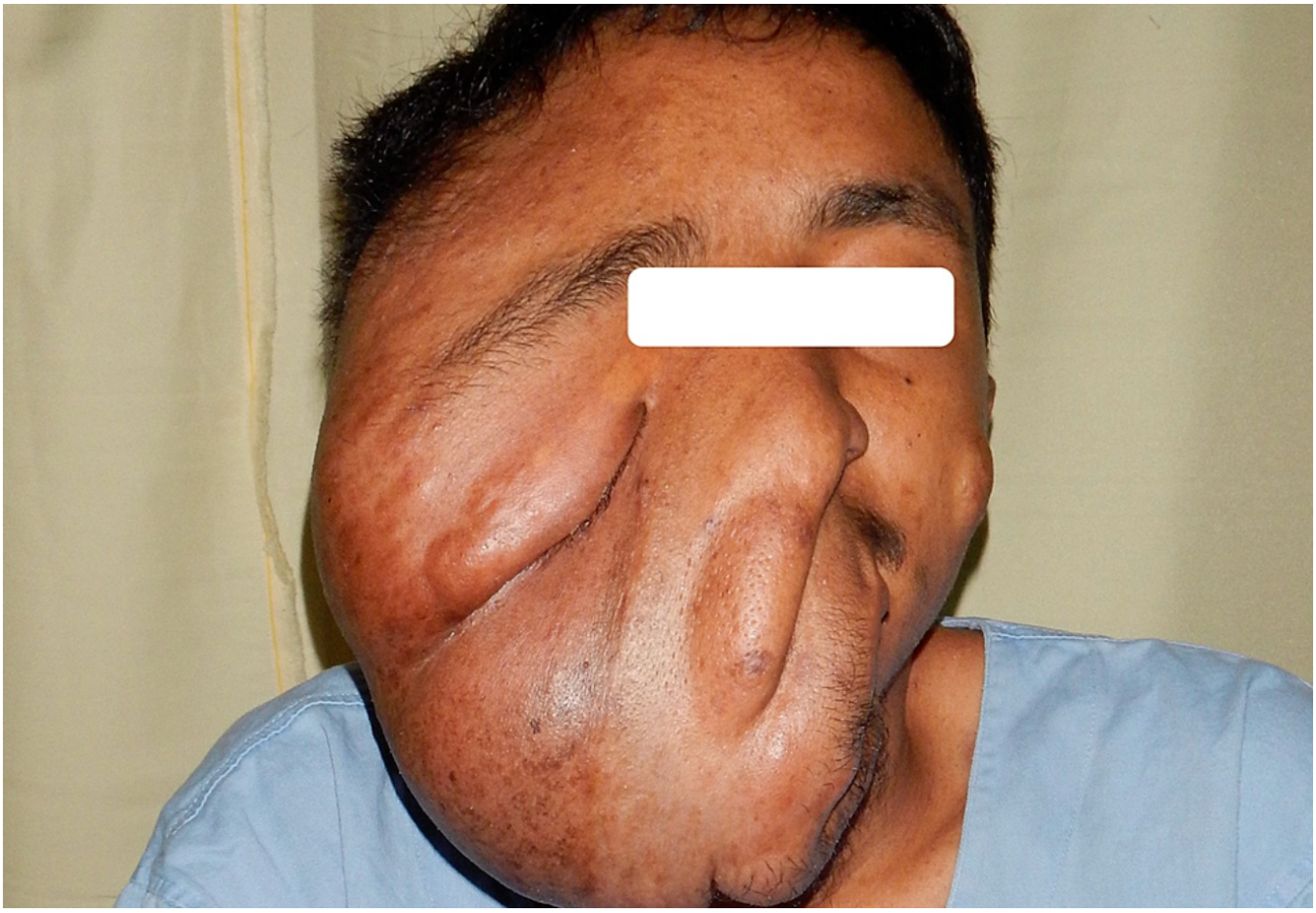
Picture Prognosis



A 14-year-old Malay male presented with a four-month history of pain and swelling on the medial side of the right knee, and a limping gait, preceded by a history of fall. Local examination showed an 8x5 cm ill-defined mass over the medial aspect of the right knee, which was hard and tender with restriction of knee joint movement. Radiographs of the right knee revealed an ill-defined lytic lesion at the proximal third of the right tibia with periosteum elevation. MRI of the right knee revealed an irregular lobulated enhancing mass measuring 4.2x4.4x6.9 cm involving the proximal metadiaphysis of the right tibia. The chest CT scan was normal. A bone scan revealed no evidence of skeletal metastasis. What's the diagnosis?

1. Hemi-Facial Plexiform Neurofibroma
2. Mesenchymal Soft Tissue Tumor
3. Lymphangioma

4. Gnatophyma

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Answer: Hemi-Facial Plexiform Neurofibroma

Neurofibromatosis type 1 (NF1) is a complex autosomal dominant, multisystem genetic disease affecting about 1 in 3500 individuals. **Plexiform neurofibromas represent a rare variant (30%) of NF1** in which the spread of tumor cells along nerve fascicles leads to a diffuse mass of thickened nerve fibers. Affected patients with NF1 have a greater chance of developing soft tissue sarcomas than the general population. Leiomyosarcoma is one of the most frequent soft tissue sarcomas, seldom observed in patients with NF1. Herein we report a rare concurrency of bone leiomyosarcoma in a patient with a plexiform neurofibroma, adding to the few reported cases