Unusual Case of Aggressive Juvenile Ossifying Fibroma Mimicking Rhabdomyosarcoma of the Masticator Space

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We present an unusual case of a 10-year-old girl presenting with a 4-month history of right ear pain progressing to right eye pain and trismus. CT showed an aggressive soft tissue tumor in the right infratemporal fossa with associated bone destruction with punctate foci of calcification and intraorbital extension. MR imaging showed a peripherally enhancing, infiltrative mass with central necrosis and intermediate T1 and heterogeneously high T2 signal centered in the right masticator space. Perfusion-weighted imaging showed elevated CBV at the periphery of the mass. The mass extended locally to involve the right medial and lateral pterygoid muscles, temporalis muscle, maxillary sinus, clivus, and sphenoid bone and sinus. The mass also surrounded the right carotid artery without compressing it, extended into the right orbit, and extended through the right foramen ovale, suggesting perineural spread along cranial nerve V. There was dural enhancement along the floor of the right middle cranial fossa without definite intracranial extension. On the basis of these imaging features, the differential diagnosis was rhabdomyosarcoma, eosinophilic granuloma, or Ewing's sarcoma. Two separate biopsies of the tumor returned a diagnosis of aggressive juvenile ossifying fibroma (JOF), with extensive spindle cell proliferation. To our knowledge, this is the first report in the English literature of aggressive JOF arising from the masticator space in a child, and only the second case in the literature of an ossifying fibroma in the masticator space.
References