ABSTRACT

Educational Objective: At the conclusion of this presentation, the participants should be able to discuss the clinical and histological features and management of masses consistent with cranial fasciitis. Objectives: The clinical history of a pediatric patient with a temporo-orbital mass consistent with cranial fasciitis is reviewed. Histology of cranial fasciitis and previously reported cases are examined to allow for comparison of this rare diagnosis. Study Design: Case report and literature review. Methods: A 2-year-old female presented with a rapidly enlarging right temporo-orbital mass that resulted in pain. MRI findings revealed erosion of the lateral orbital wall with mild compression of the globe. Results: Surgical intervention included incisional biopsy of the mass for diagnosis. Intraoperatively, the mass was found to be bright white in color and had minimal bleeding. It was well encapsulated with an obvious border. Pathological review revealed cranial fasciitis. Postoperatively, the patient did well, and a repeat MRI (approximately three months after initial scan) revealed a significant decrease in the size of the mass with almost complete resolution. No further surgical intervention was required. Conclusions: Pediatric cranial fasciitis is a rare diagnosis. Treatment may require complete surgical resection if spontaneous resolution does not occur.

CASE PRESENTATION

The patient is a healthy 2-year-old female with a two-month history of a progressively enlarging right temporo-orbital mass. There were no documented visual changes. MRI of the orbit/face/neck revealed a solid mass just lateral to the right orbit measuring approximately 2.0 cm x 2.5 cm x 2.2 cm with heterogeneous enhancement, which was more peripheral and septated. The mass appeared to destroy the lateral orbital wall and have mild extension into the lateral orbit along the lateral rectus muscle with mild impression on the globe and the lacrimal gland. It bordered anteriorly on the temporalis muscle. We performed an incisional biopsy for diagnosis. Intraoperatively, the mass was bright white in color with well-defined borders and had minimal bleeding.

RESULTS

Histologically, the tumor consisted of plump but uniform fibroblastic/myofibroblastic cells arranged in short intersecting fascicles in association with a loose myxoid matrix containing lymphocytes and red blood cells. There was no atypia or pleomorphism noted in the cells. Cytoarchitectural features consistent with nodular fasciitis, including associated bone destruction, were noted. Immunohistochemical stains showed positivity in the tumor cells for smooth muscle actin, vimentin and CD 68. Desmin, myogenin and muscle specific actin were negative. Given the clinical context, histologic appearance and immunohistochemical profile, the final diagnosis of cranial fasciitis was applied.

The patient was clinically followed very closely, and the mass began decreasing in size. Approximately 3 months after the biopsy, a follow up MRI revealed near complete resolution of the mass. No further surgical intervention was necessary.

DISCUSSION

Cranial fasciitis is a rare morphologic variant of nodular fasciitis predominantly affecting the temporoparietal region of the scalp and skull in young children (most commonly age 2-3 years), with very few reported cases occurring in adults. It is known to erode the cranium and, in some cases, the underlying dura and leptomeninges. Cranial fasciitis usually has an initial period of rapid growth followed by size stabilization around two months. Most present as a 1-3 cm subcutaneous mass; rare cases of very large masses (up to 15cm) have been reported. Current recommended treatment is surgical excision with or without curettage of the underlying bone. However, incisional biopsy has been shown in some reports to result in complete resolution. Recurrence is rare.

CONCLUSIONS

Cranial fasciitis is a rare diagnosis identified by its clinical presentation, histologic appearance, immunohistochemical profile, and radiographic characteristics. Consideration for incisional biopsy alone should precede aggressive complete surgical excision.

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REFERENCES


