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CASE REPORT

Intravascular fasciitis of the scalp as a complication of ICP monitor placement: a case report and review of the literature

Cyril S. Tankam[1] · Mason T. Stoltzfus[1] · Yaw Tachie-Baffour[1] · Julie C. Fanburg-Smith[2,3,4] · Elias B. Rizk[1]

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Abstract

Background/Importance There are only 56 documented cases of intravascular fasciitis, a rare variant of nodular fasciitis. Of these cases, only 2 involved the scalp. This lesion is amenable to surgical resection, making it important to differentiate it from soft tissue malignancies of the scalp.

Clinical presentation We report an unusual case of intravascular fasciitis involving the scalp at the site of an intracranial pressure (ICP) monitor of a 13-year-old male patient. The lesion was surgically excised with no recurrence upon 1-month follow-up.

Conclusion Intravascular fasciitis is a benign, reactive proliferation of soft tissue that may arise at sites of prior trauma. It appears as a soft, painless, mobile lesion, and immunohistochemical studies are required to differentiate it from malignant lesions. The standard of care is surgical resection of the lesion.

Keywords Intravascular fasciitis · Nodular fasciitis · Scalp

Background

In 1981, Patchefsky and Enzinger first described intravascular fasciitis (IVF) as a variant of nodular fasciitis. In their case series of 17 patients, they described a lesion that exhibited microscopic features of nodular fasciitis, but had an unusual association with veins and arteries [1]. IVF typically arises in the extremities, oral cavity, neck, and along major vessels, such as the aorta or internal jugular veins [2–30]. Only 2 cases of IVF involving the scalp have been documented in the literature [5, 31]. Here, we report an unusual case of intravascular fasciitis at the site of an ICP monitor in a patient's scalp. We also present a review of the literature pertinent to this case of intravascular fasciitis.

✉ Mason T. Stoltzfus
mstoltzfus3@pennstatehealth.psu.edu

[1] Department of Neurosurgery, Penn State Children's Hospital, PA, Hershey 17033, USA

[2] Department of Pathology, Penn State Children's Hospital, Hershey, PA, USA

[3] Department of Pediatrics, Penn State Children's Hospital, Hershey, PA, USA

[4] Department of Orthopedics, Penn State Children's Hospital, Hershey, PA, USA

Case presentation

A 13-year-old male with a history of Chiari malformation, myelomeningocele status post repair, and ventriculomegaly, status post insertion of a ventriculoperitoneal shunt, presented for a follow-up 71 days after placement of a Codman CereLink ICP sensor and was found to have swelling at the incision site of the monitor. He reported no headache, nausea, vomiting, seizure, change in behavior, or cognitive decline. An X-ray of the skull was negative for retained hardware. An ultrasound of the area revealed a 1.5×1.5 cm hypodensity. Fine needle aspiration retrieved approximately 2 mL of blood, mildly reducing the swelling. After consulting with the patient and their parents, the decision was made to excise the mass. The patient and his parents consented to this procedure. Excision of the mass revealed a well-demarcated, soft, compressible, blue-purple spherical lesion with an intact capsule and adhesion to the underlying skull. The specimen was sent for pathological examination. Histopathologic examination revealed a vessel distended by spindled tissue culture like myofibroblasts, focal pericytoid cells and neovessels, keloidal collagen, and myxoid degeneration with extravasated erythrocytes and lymphocytes that resembled an intravascular organizing thrombus and classified as intravascular fasciitis (Fig. 1). The patient

tolerated the procedure well and there were no complications. Upon 1-month follow-up, inspection of the surgical site revealed a well-healing incision with no tenderness or residual swelling.

Discussion

Nodular fasciitis is a benign soft tissue proliferation involving fibroblasts and myofibroblasts that typically arises from subcutaneous tissue, fascia, or muscles. Intravascular fasciitis is a rare type of nodular fasciitis; only 56 cases have been documented in the literature, with only 2 involving the scalp [2, 5, 31]. This lesion arises as a reactive proliferation of myofibroblast inside the vascular lumen or in the fascia investing small- and medium-sized blood vessels [25].

This form of nodular fasciitis typically arises in young adult patients with a preference for the head, neck, and extremities [1, 4, 10, 13, 23, 24, 30, 33–35]. When it arises on the skin, it is typically a painless, mobile mass often brought to attention for cosmesis, rather than due to physical discomfort. These lesions are typically solitary nodules, but may present as multiple or multinodular masses. In some cases, IVF arises directly from the veins (external jugular vein, femoral vein, etc.) and major arteries (aorta) [3, 17, 20, 27, 36]. In these cases, it commonly presents with symptoms of venous thrombosis or aortic dissection [3, 5, 7, 18, 22].

Microscopically, IVF is characterized by spindle cells arranged in intersecting fascicles, typically on a fibrous, vascular, or myxoid background. Mitotic figures tend to be rare or absent. Nuclear atypia is rarely seen. Immunohistochemical staining shows that the spindle cells typically stain positive for vimentin and alpha-smooth actin and negative for keratin, S100, desmin, CD31, CD34, and c-kit, indicating they are derived from myofibroblasts [10, 16, 18, 19, 23, 26, 32].

Because it tends to involve blood vessels, intravascular fasciitis can be confused with malignancy, such as fibroblastoma, myofibroblastoma, fibrosarcoma, leiomyosarcoma, or liposarcoma [12, 14, 37–39]. In some cases, lesions of intravascular fasciitis exhibit atypical changes such as increased mitotic activity. Despite these changes, these lesions rarely resemble malignancy through metastasis or recurrence [12].

So far, the pathogenesis of intravascular fasciitis is likely related to trauma and with a USP6 fusion. As in our case, these lesions can arise at sites of prior trauma. Other possible predisposing factors include thrombosis (specialized organizing thrombus) or pregnancy-related changes [9, 16, 21, 34, 38, 40].

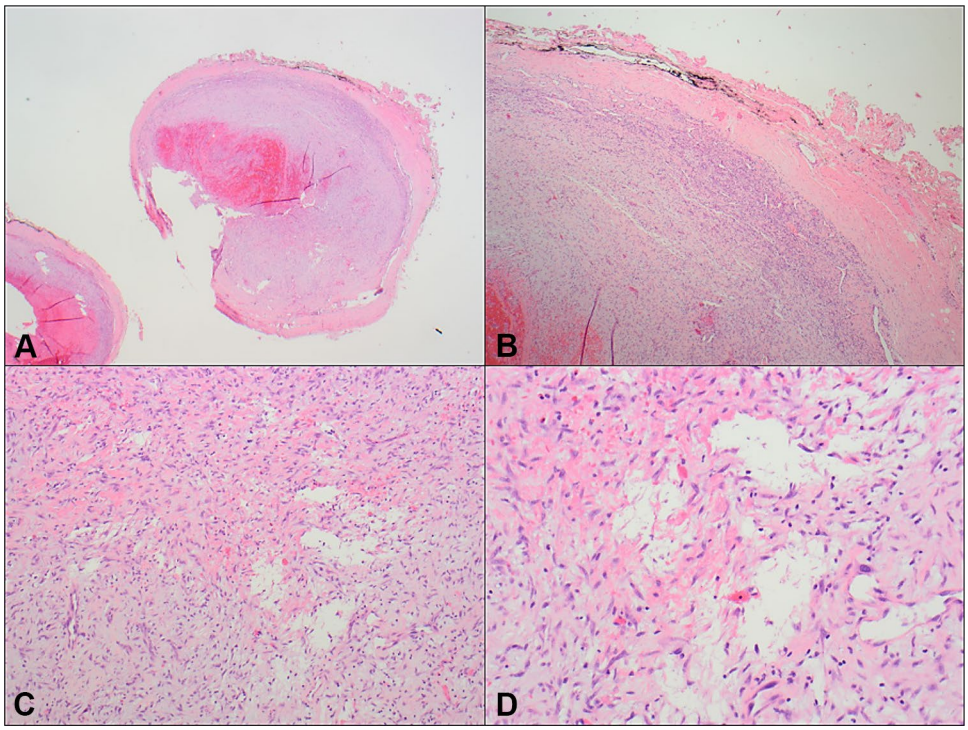

Fig. 1 Histopathology of the scalp intravascular fasciitis: low power demonstrating cross section of a distended vessel filled with organizing thrombus-like spindled tissue-culture like myofibroblasts (**A**–**D**), pericytoid vessels (**B**), and keloidal collagen and myxoid degeneration with extravasated erythrocytes and lymphocytes (**C**, **D**)

Conclusion

Intravascular fasciitis of the scalp is uncommon. It can be misidentified as benign lesions, such as local thrombosis at a site of injury, or more serious conditions, such as sarcoma or mesenchymal neoplasms, that share its characteristic rapid growth. Immunohistochemistry, gross appearance, and clinical symptoms can be used to differentiate intravascular fasciitis from these lesions. Surgical excision should be the standard of care for these lesions, as rates of recurrence are very low.

Author contribution Cyril Tankam and Yaw Tachie-Baffour performed the literature review, prepared the figures, and wrote the main manuscript. Yaw Tachie-Baffour, Elias Rizk, Mason Stoltzfus, and Julie Fanburg-Smith assisted in the literature review and writing of the manuscript. All authors reviewed the manuscript and approved the document for submission.

Data availability All the material is owned by the authors and/or no permissions are required.

Declarations

Ethics approval and consent to participate Not applicable since no identifying images or other personal or clinical details of participants that compromise anonymity were included in this manuscript; therefore, no consent was obtained for publication.

Consent for publication The authors consent that this document should be published.

Conflict of interest I declare that the authors have no competing interests as defined by Springer, or other interests that might be perceived to influence the results and/or discussion reported in this paper**.**

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