Adult periorbital fibromatosis

CATHERINE N SMOOT, GREGORY B KROHEL, AND RICHARD S SMITH

From the Department of Ophthalmology, Albany Medical College, New York

SUMMARY Three patients aged 15 to 40 years with fibromatosis of the periorbital region were studied. A history of acute onset, preceding trauma, and pain associated with a palpable mass were typical features. Rapid recurrence followed surgical excision. Misdiagnosis is common in this benign disease and may lead to unnecessary examinations for malignancy. Surgical excision or debulking of these masses is indicated for diagnosis and pain relief, but long-term follow-up indicates that these lesions will often spontaneously resolve with time.

Benign acute recurrent fibromatosis is a fibroblastic proliferation that lies in a range between normal repair with scar formation and fibrosarcoma. Reports have suggested that trauma may play a part in the development of this disease,¹ though other reports dispute this connection.^{2,4} These fibrous masses have no definable margins and may locally invade bone and striated muscle. They do not tend to metastasise.^{3,5,6} The pathological diagnosis may be confusing and malignancy is often initially suspected.

Juvenile fibromatosis is a well described clinical entity that occurs throughout the body, though the abdomen appears to be the most frequent site.¹ Fibromatosis of the periorbital region has previously been described in children.⁴ Clinical characterisation of this entity in adults has not been described in the ophthalmic literature. We recently treated three women with recurrent periorbital fibromatosis.

Case reports

A 24-year-old nursing student first presented to Albany Medical Center Hospital (AMCH) in January 1984 with a corneal ulcer refractory to treatment. She developed ecchymosis, swelling, and warmth of the left infraorbital region while in the hospital. This appeared to respond to intravenous antibiotic therapy, though a small nodule remained over the left premalar area. In April the patient was noted to have increasing ecchymosis of the left lower lid and premalar region. An excisional biopsy via a subciliary eyelid incision revealed diffuse fibrotic tissue over the premalar area and extending into periosteum. The

initial pathological interpretation was rhabdomyosarcoma and an oncology specialist was consulted. Re-examination of the specimen suggested a reactive myositis.

One month later the patient presented with a painful ecchymotic mass and a small eschar in the premalar region. A second biopsy was obtained and a diagnosis of fibromatosis was made. In October of 1984 the eschar enlarged and she underwent drainage of an abscess over the left malar eminence. Manipulation of the wound site by the patient was suspected and she was referred to the psychiatric service for evaluation of probable Munchausen's syndrome.⁷ Her clinical course improved slowly under psychiatric observation. No recurrence of her problem has been noted over the past two years.

CASE 2

A 15-year-old female presented to AMCH in May 1981 with a history of spontaneous left malar and infraorbital ecchymosis. It was later thought she might have suffered injury from her boyfriend. A painful, palpable, ecchymotic mass was noted along the left inferior rim (Fig. 1). A subciliary excisional biopsy was performed and initially interpreted as nodular fasciitis. Further review and consultation with the Armed Forces Institute of Pathology confirmed the diagnosis of benign fibromatosis. The patient experienced several recurrent episodes of painful ecchymosis, with regrowth of the mass. A repeat anterior orbitotomy performed six months later confirmed the diagnosis of fibromatosis. Over the next three years she had several episodes of painful swelling and ecchymosis lasting one to two weeks. Each recurrence tended to be less severe and resolved spontaneously. Further surgery was not necessary.

Correspondence to Gregory B Krohel, MD, Albany Medical College, Department of Ophthalmology, Albany, New York 12208, USA.

Fig. 1 Ecchymotic mass over the left infraorbital and malar area secondary to fibromatosis.

CASE 3

A 40-year-old nurse was accidentally struck forcefully in the left infraorbital area by a patient's head in November 1986. This resulted in persistent, painful infraorbital swelling over the left malar area accompanied by a dimpled contracture. Two months later a fibrotic mass was removed by anterior orbitotomy through a subciliary incision. Pathological examination confirmed the clinical diagnosis of fibromatosis. Two weeks postoperatively she presented with a recurrent painful mass. The decision was made to institute massage therapy with vitamin E cream while observing the clinical course. The dimpling and pain had decreased four months later. A small pea sized nodule remained. By September 1987 the dimpling and pain had resolved, and only a small soft nodule over the left lateral premalar area remained. No further surgery is planned.

Discussion

Fibromatosis in adults and children has been previously described in various sites of the body including the periorbital region.^{1,3,4,8,9} Our three cases of adult periorbital fibromatosis had several features

in common. Two patients had a definite history of trauma immediately preceding the onset of the lesion and trauma was strongly suggested in the third case. Trauma has been mentioned as an aetiological agent in some cases of fibromatosis,¹ though five of six patients previously described with juvenile periorbital fibromatosis had no history of trauma.⁴ Despite the ecchymotic appearance of these lesions haemosiderin was observed in only one of our three patients.

All three patients in our series were women. A female preponderance had been noted previously, suggesting a hormonal influence in the development of fibromatosis.⁸

The prevalence of pain with fibromatosis remains uncertain. Conley *et al.*³ noted an absence of pain in seven patients with periorbital lesions, while two patients with periorbital lesions in another series had associated pain.⁸ The onset of symptoms was acute and painful in the three patients we treated. This contrasts with the cases of juvenile periorbital fibromatosis previously reported in the ophthalmic literature which were described as firm, painless lumps, with no mention of acute onset.⁴

Misdiagnosis of this benign tumour can occur.⁹ One of our patients underwent examination for suspected rhabdomyosarcoma. The other misdiagnoses in our patients were reactive myositis and nodular fasciitis.

A surgical approach via a subciliary incision in all three patients yielded excellent postoperative cosmesis (Fig. 2). A firm, whitish rubbery mass extending from subcutaneous tissue to periosteum was observed in all cases. Integrity of the surrounding

Fig. 2 Postoperative appearance of patient in Fig. 1 with a good cosmetic result. The patient had several recurrences following surgery.

Fig. 3 A focus of perivascular fibrous tissue with lymphocytic infiltration (arrow) consistent with fibromatosis. (Haematoxylin and eosin $\times 150$).

Fig. 4 Intracellular haemosiderin deposits are present (arrows). Haemosiderin was noted in only one of three patients despite the ecchymotic appearance of all three lesions. (Haematoxylin and eosin, $\times 60$).

bony structures was preserved in two cases, though local bony invasion similar to that previously reported was observed in one patient.³

Histopathologically, fibromatosis is a proliferation of fibroblasts with interstitial collagen and rare mitotic activity.⁹ This differs from fibrosarcoma which has less interstitial collagen and more mitotic activity.¹⁰ Hidayat and Font⁴ microscopically noted a leiomyomatous appearance of the tumours in three of the juvenile patients they reported, a picture similar to that of congenital generalised fibromatosis. Our pathology sections showed dense perivascular collections of fibrous tissue with frequent foci of lymphocytes in all specimens (Fig. 3). The fibroblastic cells varied in shape from spindle to stellate, while nuclei differed in their size and degree of basophilic staining. Haemosiderin was noted in the pathological specimens in one case (Fig. 4). Electron micrographic (EM) studies confirmed the fibroblastic nature of the lesion in one patient. Characteristic features shown included a proliferation of mesenchymal fibroblasts with an increase in rough cytoplasmic reticulum and the presence of intercellular fibrin and collagen.

The treatment employed was local surgical excision of the lesion. The therapeutic value of steroids,^{6,11} radiation,^{3,12} or chemotherapy¹³ remains inconclusive. Despite good surgical results all our patients had recurrences. We decided to reoperate only if the pain could not be controlled medically. Two of our three patients required further surgery for that reason, while one was successfully managed with time and massage after the initial resection.

In summary, benign acute idiopathic fibromatosis should be considered in the differential diagnosis of

acute, tender, palpable periorbital lesions in adults. A careful history may reveal a preceding traumatic incident. Surgical excision affords local pain relief and cosmetic improvement, but recurrences are common.

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