Spontaneous recurrent hemorrhage as an unusual complication of fibrous dysplasia of the skull

Case report

CARL J. GRAF, M.D., AND GEORGE E. PERRET, M.D.

Division of Neurosurgery, Department of Surgery, University of Iowa Hospitals and Clinics, Iowa City, Iowa

The case of a patient with fibrous dysplasia of the skull associated with recurrent spontaneous hemorrhage is presented. Suggestions for management of this unusual complication are made.

KEY WORDS • fibrous dysplasia • spontaneous hemorrhage

Fibrous dysplasia of the skull manifests itself as a local deformity. When it involves the cranial vault, the deformity is usually one of a hard, sometimes tender, prominence. Commonly the lesion presents at the fronto-orbital region and involves the frontal, sphenoid, and maxillary bones, resulting in facial asymmetry. Proptosis may be associated with diplopia; visual loss may occur from compression of the optic nerve by bone. More extensive involvement may include the ethmoid, nasal, temporal, or zygomatic bones.

The purpose of this report is to record the unusual complication of recurrent spontaneous hemorrhage in an area of fibrous dysplasia.

Case Report

This 9-year-old girl was examined at the University of Iowa Hospitals in March, 1967. For 3 months she had had a painless lump at the left frontotemporal region and swelling of the forehead above the saddle area of the nose. Skull films disclosed increased density of the greater and lesser wings of the left sphenoid, left zygomatic, and left frontal bones, and orbital roof. The left ethmoid air cells were obliterated. The left antrum was barely aerated, being replaced by dense bone. A diagnosis of fibrous dysplasia was made (Fig. 1). During the next 2 years, no further changes were noted in the physical or x-ray examinations of the skull. In 1972, at the age of 14 years, the patient had the acute onset of pain over the left frontal region. The pain persisted for 3 or 4 days, followed by progressive swelling of this area, after which the pain became less pronounced. There was no history of injury to the head. Examination disclosed a 5 x 5 x 2 cm soft, cystic, nontender, nonpulsatile mass over the left frontal region; there was no bruit and no change in color of the overlying skin. The mass did not transilluminatet. X-ray films revealed that the dysplastic process had spread into the left parietal bone, and was characterized by irregular areas of radiolucency interlaced with areas of thickened bone. There was expansion of the outer table of the left frontal bone, irregularity of the inner table, and a large soft-tissue swelling overlay a large lytic bone defect (Fig. 2). After aspiration of 60 cc of thick, grossly bloody fluid, the area became flat. A pressure dressing was applied. On the following day, a ledge of bone was palpable surrounding a depressed central area 5 cm in diameter.

At the age of 17 years (November, 1975), the patient noted that the left frontal region was more depressed, as with further loss of bone. The left eyeball was prominent. X-ray films showed greater involvement of bone, with cystic changes that included the right frontal bone, the left orbit, and the left zygoma.

In June, 1977, the patient reported that she had suffered intermittent left frontal pain of 3 months’ duration. Films of the skull revealed more extensive changes than in 1975, involving in particular the left
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FIG. 2. Tangential film outlining destruction of frontal bone with overlying soft-tissue swelling representing subgaleal hematoma.

zygomatic maxillary region. In November, 1977, although there was no increase in prominence of the left frontal bone, the patient complained of pain and tenderness at this site. In December, 1978, progressive headache and greater prominence of the left and midfrontal area were noted; the region became tender and fluctuant. A diagnosis of spontaneous hemorrhage was made, and, although aspiration was recommended to confirm the impression, this was refused by the patient.

In the next 3 weeks the swelling became considerably greater. Examination showed a round, tender, soft, fluctuant swelling, 10 × 10 × 5 cm in size, over the anterior mid and left frontal vertex of the head. There was no bruit or pulsation. Left exophthalmos and prominent protrusion of the left supraorbital, infraorbital, and zygomatic regions were noted. We aspirated 115 cc of blood from beneath the scalp, but within 1 to 2 minutes the degree of fullness was as great as before, so that attempts to apply a pressure dressing were futile. On the following day, despite aspiration at different depths and sites, only 40 cc of dark blood could be obtained, and although the hematoma cavity became less tense, it did not collapse completely. The aspirated material contained 1,630,000 red blood cells, a hemoglobin value of 5.7 gm, and a hematocrit of 16%.

On January 4, 1979, because of the fear of a bleeding arteriovenous malformation or disrupted bleeding diploic channels, left common and left selective external carotid angiography was performed. Immediately before angiography, 50 cc of material, grossly indistinguishable from pure blood, was again aspirated. The mass collapsed almost completely, but within 5 minutes it was as tense and full as before aspiration. The angiogram showed no vascular malformation or increased vascularity of either the bone or overlying soft tissue. Two terminal branches of the superficial temporal artery ran to the involved area, but there was no disruption of the vessels to indicate that they were the source of bleeding (Fig. 3). A technetium-99 dynamic scan showed uptake of the nuclide in the periphery of the left frontal bone lesion in the venous phase, whereas the central portion

FIG. 1. Roentgenogram showing eburnation of bone involving the frontal and lesser wing of the sphenoid bones (arrows).

FIG. 3. Left external carotid angiogram showing terminal branches of the superficial temporal artery at the defect in the frontal bone.
FIG. 4. *Left:* Dynamic brain scans showing uptake of nuclide in the peripheral portion of the bone lesion (arrow). *Center and Right:* Diffuse uptake of nuclide in the bone lesion.

showed no activity. At 3 hours, the static scan showed greater activity in a peripheral rim, and uptake of the central part of the lesion as well (Fig. 4). On January 15, 1979, the mass was smaller, measuring $4 \times 4 \times 2\frac{1}{2}$ cm, and irregular bone edges were palpable beneath it. Some 10 days later the scalp was flat. Examination on February 29, 1979, showed an excavation, $6 \times 5 \times 1$ cm in area, in the left frontal region of the skull; the base was firm (Fig. 5).

**Discussion**

The pathology of fibrous dysplasia is one of new bone formation within which dense sheets of fibrous tissue are laid down. The bony trabeculae are surrounded by osteoblasts, osteoclasts, and osteoid tissue. Jelsma stated that the lesion is more vascular than is normal bone. Feiring, *et al.*, in discussing five patients upon whom they operated and who were found to have fibrous dysplasia, made no mention of excessive hemorrhage or vascularity during operation. In one instance, they stated "the tissue was of uniform consistency and devoid of large blood vessels," whereas in two others they mentioned occasional areas of blood associated with giant cells when describing the histopathology of the operative specimens. Matson described a "unique indication for surgery in a 14-year-old girl who had recurrent exsanguinating nosebleeds which required radical removal of involved paranasal and intracranial bone together with bilateral external carotid artery ligation."

Sarcomatous change may rarely occur in areas of fibrous dysplasia. Long, *et al.*, stated that if an "active increase in size or local inflammation becomes apparent, then total excision of the lesion with cranioplasty is the treatment of choice." They indicated that "the area of fibrous dysplasia is very vascular and blood loss during such a procedure is likely to be considerable." Our own operative experiences with fibrous dysplasia have not impressed us by unusual problems with hemorrhage. Despite these
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Figure 5. Lateral (upper) and frontal (lower) roentgenograms showing extensive lesion with excavation of frontal bone. There is no overlying soft-tissue swelling.

remarks on the highly vascular nature of the lesion, the only reference we found regarding spontaneous hemorrhage is that described by Matson. No mention is made as to whether bleeding in his patient was arterial or venous, but the description suggests that it was of arterial nature.

We believe that the bleeding in our patient occurred from open diploic channels resulting from breakdown of bone in the area of fibrous dysplasia. We believe that the bleeding was of venous nature and produced rapid distention of the scalp overlying the outer table of the skull that was being destroyed. In our patient, very rapid bleeding took place after the hematoma was evacuated by aspiration, suggesting that open diploë were the source of the hemorrhage. It is likely that no further accumulation of blood occurred when the pressure of the mass of blood became equal to that inside the diploë. Clotting later took place, followed by resorption of blood and disappearance of the swelling.

The rapid increase in size of the mass raised the question of malignant (sarcomatous) change in the lesion. Our experience in this case should alert one, as well, to the possibility of the lesion producing a benign hemorrhage beneath the scalp and that it might better be left alone unless the involved scalp becomes compromised, as evident from inflammatory changes or the appearance of solid tissue to indicate new growth. The risk of introducing a serious infection by an aspirating needle is unnecessary because the hematoma will resolve spontaneously. Furthermore, to attack it surgically in the “acute” bleeding stage may lead to serious blood loss necessitating extensive bone resection, which may be incomplete and still not solve the problem.

References


Address reprint requests to: Carl J. Graf, M.D., Division of Neurosurgery, University Hospitals, Iowa City, Iowa 52242.