Radionuclide Angiography—Brain and Bone Imaging in Craniofacial Fibrous Dysplasia (CFD): Case Report

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Craniofacial fibrous dysplasia (CFD) may show arterial displacement or increased uptake during arterial, capillary, venous, and blood-pool phases of radionuclide angiography. Extensive single or multiple areas of uptake are present on brain and bone images. These findings are probably due to the highly vascular bone often found in CFD. The distribution of the areas of abnormal uptake, along with pertinent clinical history and skull radiographs, usually allows other conditions to be excluded.

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Although examples of static brain images in craniofacial fibrous dysplasia (CFD) have been published, the finding on radionuclide angiography and bone imaging are not well described (*1–2*). This report notes the findings from [^{99m}Tc] sodium pertechnetate angiography, blood-pool and static brain imaging, and Tc-99m polyphosphate bone imaging in three patients with CFD.

CASE REPORTS

Case 1. A 26-year-old woman was admitted with decreasing vision in the left eye. The clinical diagnosis of CFD had been made 5 years earlier. Skull radiographs on admission showed sclerosis and expansion of the left frontal, parietal, sphenoid, and ethmoid bones; the right frontal and parietal bones were also involved (Fig. 1A). Pertechnetate angiography showed medial displacement of the left middle cerebral artery; there was increased uptake on the left side anteriorly in the venous and blood-pool phases (Fig. 1B). The static brain images 30 min later showed intense uptake on the left side anteriorly (Fig. 1C). The Tc-99m polyphosphate bone images showed involvement of the facial bones on the left, and of the calvarium bilaterally (Fig. 1D). The left mandible also showed intense uptake. No other involvement of the skeleton was noted. The serum alkaline phosphatase was normal. Left orbital decompression was planned should the loss of vision progress.

Case 2. A 42-year-old woman presented with

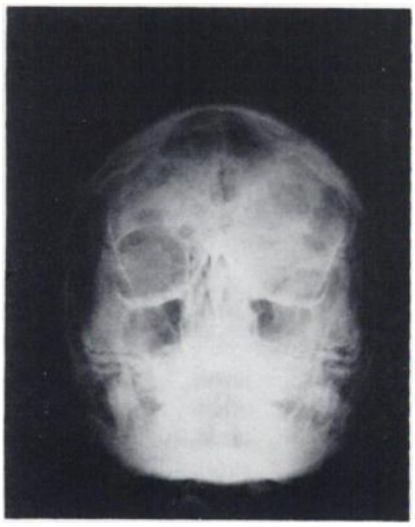

FIG. 1A. Case 1. Water's view: note marked sclerosis of craniofacial bones on left, with extensive involvement of left orbit.

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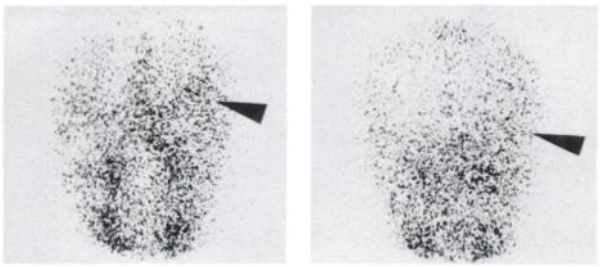

FIG. 1B. [^{99m}Tc] Sodium pertechnetate angiogram, arterial phase: anterior view shows medial displacement of left middle cerebral artery (left). Increased uptake in left anterior craniofacial bones is noted on the anterior views of venous and blood-pool phases (right).

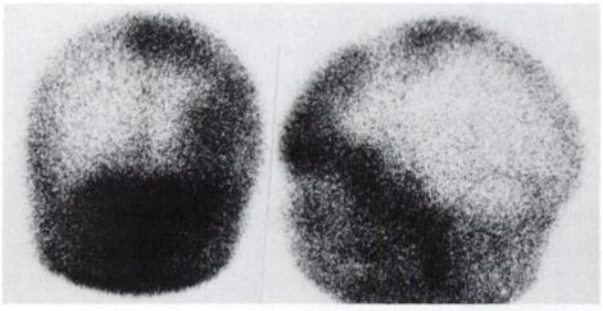

FIG. 1C. Pertechnetate static images: anterior and left lateral views at 30 min after injection show extensive uptake on left anteriorly in same area as radiographic abnormality.

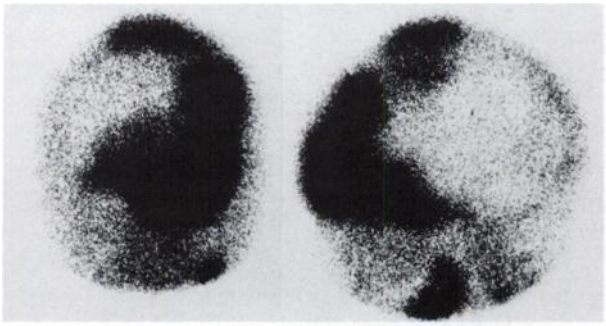

FIG. 1D. Tc-99m polyphosphate images: anterior and left lateral views show more extensive involvement of right craniofacial bones including mandible.

proptosis of the right eye of 7-years duration. Hyperthyroidism had been diagnosed shortly before the onset of proptosis, and radioiodine therapy was given. It was thought that the unilateral proptosis was a manifestation of Graves' disease. Skull radiographs were not made at that time. She was admitted now for consideration of cosmetic correction of the proptosis. Skull radiographs revealed sclerosis, primarily of the right sphenoid and maxillary bones (Fig. 2A). Pertechnetate angiography showed increased uptake in the right frontal region during the arterial, venous, and blood-pool phases (Fig. 2B). Static brain images 30 min later showed increased uptake at the right base anteriorly (Fig. 2C). Subsequent Tc-99m polyphosphate bone imaging showed intense uptake in the same area (Fig. 2D). No abnormal extracranial skeletal uptake was present. A left common carotid arteriogram showed no abnormal skull or intracranial vascularity. Because of the findings from brain and bone imaging, the relatively early age of onset, and a normal serum alkaline phosphatase, the diagnosis of CFD was made. Cosmetic surgery was not performed.

Case 3. A 40-year-old man was admitted with unilateral proptosis of the left eye of 15-years duration. The clinical diagnosis of CFD had been made many years before. There had been questionable progression of the proptosis over the 2 years before admission. Skull radiographs showed sclerosis of the left frontal bone (Fig. 3A). A radionuclide angiogram was unsuccessful. Static pertechnetate brain images showed increased uptake at the left frontal

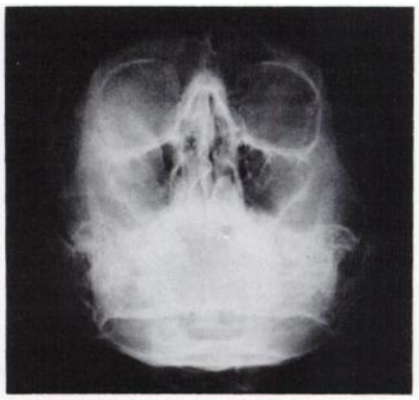

FIG. 2A. Case 2. Water's view: sclerotic involvement of right orbit is evident.

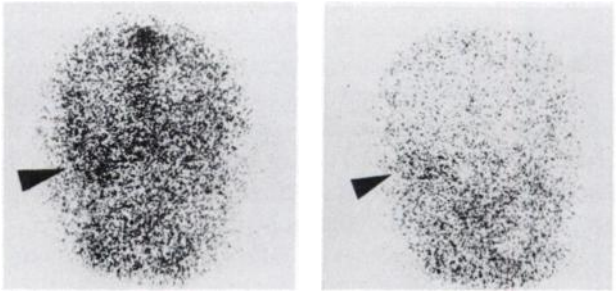

FIG. 2B. Pertechnetate angiogram, anterior view: arterial capillary (6–8 sec) and late venous (14–16 sec) and blood pool phases show increased uptake anteriorly in region of sclerotic bone on Water's view.

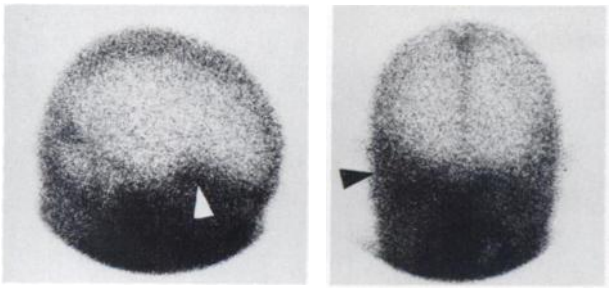

FIG. 2C. Pertechnetate static images: anterior and right lateral views 30 min after injection show increased uptake in area of sphenoid bone (arrow).

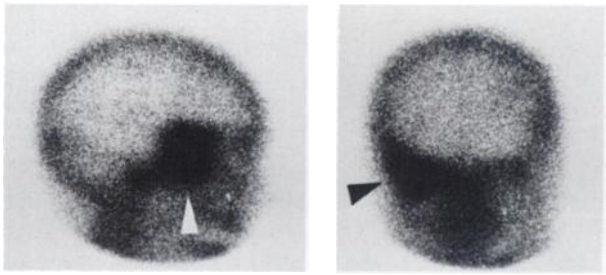

FIG. 2D. Tc-99m polyphosphate images: anterior and right lateral views show more extensive involvement of sphenoid and posterior part of maxilla (arrow).

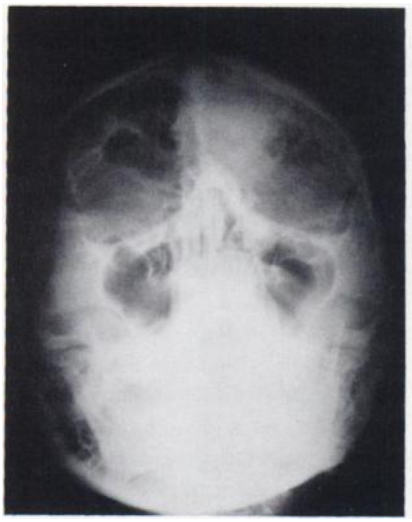

FIG. 3A. Case 3. Water's view: sclerotic involvement of left frontal and ethmoid bones is apparent.

area (Fig. 3B). The Tc-99m polyphosphate bone images showed intense uptake in the left frontal bone (Fig. 3C); the skeleton was otherwise normal. The serum alkaline phosphatase was normal. A left orbital decompression was performed.

DISCUSSION

Craniofacial fibrous dysplasia often presents during late childhood or young adulthood, and is more common in females (*3*). Up to 27% of patients with CFD have at least one other site of skeletal involvement (*4*). The frontal, sphenoid, and occipital bones seem most commonly involved (*4–5*). Skull asymmetry is the most frequent presenting complaint (*4*). The disease often becomes quiescent during adulthood (*5*). Histologically the lesion is characterized by a cellular proliferation of fibroblastic tissue in a swirling pattern, with scattered bony or osteoid trabeculae (*6*). Radiographically the bone may appear lucent or sclerotic depending on the degree of ossification present. The appearance presents a spectrum from a cystic lucent lesion to a dense, sclerotic, expanding lesion suggesting Paget's disease (*7*).

The bone of fibrous dysplasia may be highly vascular (*8–9*). This vascularity probably accounts for the increased uptake during pertechnetate dynamic and static brain imaging, and Tc-99m polyphosphate bone imaging. The pattern might be confused with that of a variety of vascular tumors, or an arteriovenous malformation (*10,11*), or a cerebrovascular accident with the "luxury perfusion syndrome" (*12*), or Paget's disease (*13*). The characteristic findings on skull radiographs would tend to rule out most lesions except meningioma en plaque, Paget's disease, neurofibromatosis, and osteoblastic metastases,

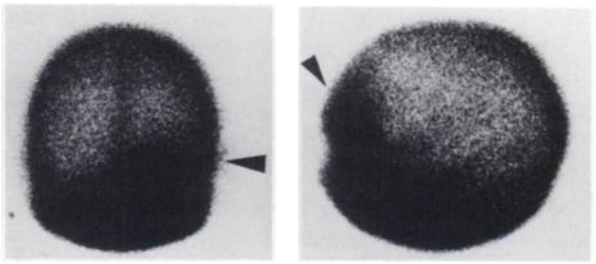

FIG. 3B. Pertechnetate static images: anterior and left lateral views 30 min after injection show left anterior frontal uptake (arrow).

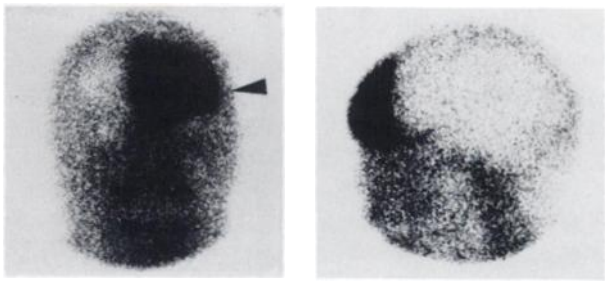

FIG. 3C. Tc-99m polyphosphate images: anterior and left lateral views show intense extensive left frontal uptake (arrow).

all of which may mimic localized fibrous dysplasia (*4,13*). Abnormal vascular supply from the external carotid system may be seen on contrast angiography in meningioma en plaque, but biopsy may be necessary to establish a definite diagnosis. The finding of multiple areas of uptake on bone imaging, as in Case 1 (Fig. 1D), would be most unusual in meningioma en plaque. The single extensive area of uptake shown in Case 3 (Fig. 3C), and the extensive uptake in the sphenoid and posterior part of the adjacent maxilla in Case 2 (Fig. 2D), would also be uncommon in meningioma en plaque. Meningioma en plaque tends to involve smaller, quite circumscribed areas with hyperostosis of the inner table rather than the entire bone (*4*).

Paget's disease may be radiographically quite similar to CFD, but the age of onset is usually later (*14*). The serum alkaline phosphatase level is almost always elevated in Paget's disease (*14*), but is often normal in adults with CFD (*4*). A normal serum alkaline phosphatase level would tend to exclude the possibility of osteoblastic metastasis. Patients with neurofibromatosis often present other manifestations of the disease (*15,16*).

Contrast angiography may reveal abnormal displacement or vascularity of the anterior or middle cerebral vessels, but is most often normal in CFD (*8*). Case 1 showed upward and medial displacement of the left middle cerebral artery during nuclide angiography, and increased uptake in the static brain images. This arterial displacement might be confused with a localized subdural hematoma (*17*), but the increased uptake during the blood-pool phase, the striking uptake in multiple areas on bone imaging, and the characteristic skull radiographs, allow this diagnosis to be properly excluded. Case 2 had conventional contrast angiography; subtraction films did not show any abnormal vascularity, but increased vascularity of the bone is strongly suggested by the increased uptake on nuclide imaging of the blood pool.

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