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Fibrodysplasia of the Skull: Case Report and Review of the Literature

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CASE STUDY

The patient is a 66 year-old white female who one month prior to admission coughed and felt a pain in the waistline area. She went to her family physician who ordered an x-ray. She also had a bone scan and was found to have a lytic lesion in the skull. She was referred for a neurosurgical consultation. Bone scan also showed focal increased activity at the T-12 area consistent with compression fracture of relatively recent origin. CT scan of the left temporal bone showed a 2.2 x 0.7 cm diameter lytic lesion in the left posterior parietal bone. Past history was significant only for a motor vehicle accident about one year prior in which she was rear-ended resulting in minor injuries. Neurological evaluation did not show any focal neurological deficits. Metastatic work-up did not reveal any evidence of primary lesion. She underwent a left parietal skull biopsy and removal of the lesion with cranioplasty. The pathology was consistent with fibrodysplasia of the skull.

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Fibrodysplasia is a benign developmental anomaly resulting in defective endochondral bone maturation. There is no evidence of a Mendelian pattern of inheritance.[7] It begins in the skeletally mature patient but age at onset of symptoms varies widely. In general, fibrodysplasia clinically manifests in late childhood. Most patients present with pain, but many will remain asymptomatic into adulthood.[8]

The sclerotic and cystic forms appear in younger patients (20 years of age) versus the mixed form which usually presents in patients 30 years of

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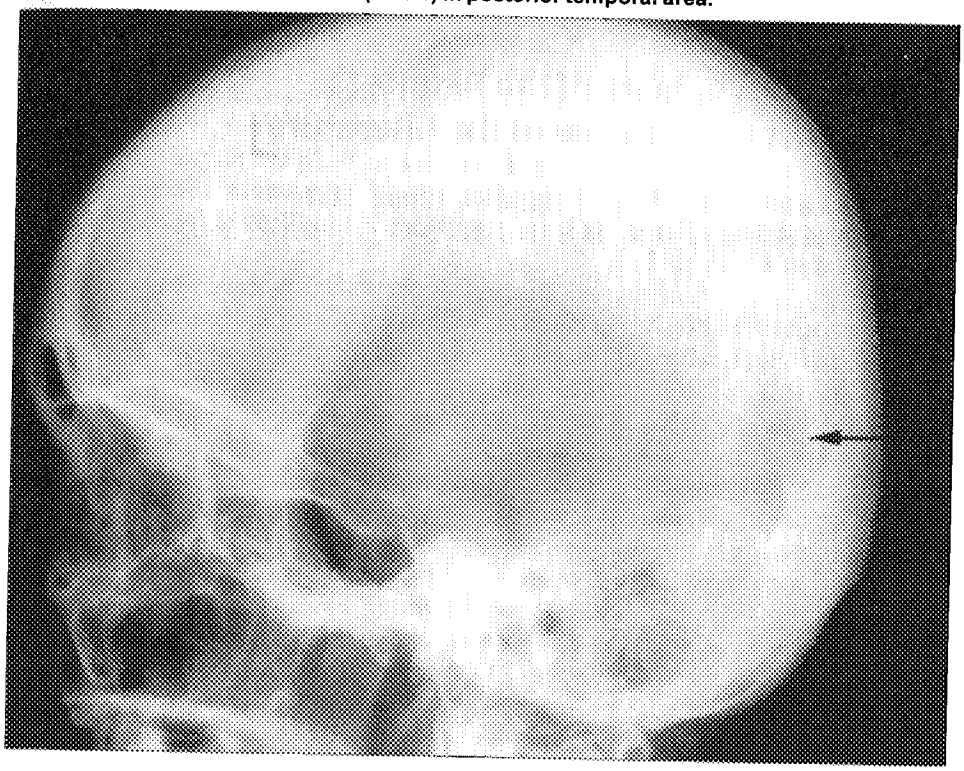
Figure 1. Lytic lesion of the skull (arrow) in posterior temporal area.



age and older. This implies that the mixed form evolves from the sclerotic and cystic lesions. Swelling and localized tenderness may be observed over the affected area. In addition to the disfiguration of the skull in the polyostotic form, symptoms of the monostotic form of the disease include headaches, convulsions, exophthalmos, optic atrophy, and deafness.[6,8]

Often, the disorder becomes clinically apparent after minor trauma.[2,3] Symptoms may begin at any age, but onset usually occurs in early adult life. Family history is negative, and there is no racial or sexual predominance.[8]

Fibrodysplasia of the skull radiologically represents in the following different types.

1) Cystic type: radiolucent center bulging outward with no protrusion inward.
2) Sclerotic type: increased bone density mimicking meningiomas.
3) The mixed sclerotic and radiolucent type.[10]

Most of the lesions that occur in the skull are of the sclerotic variety.[11] Radiologic occurrences are variable and depend upon the amount of bone laid down in the affected areas.[2,3,5,6]

Surgical treatment is indicated if important functions are threatened, there is evidence of severe deformity, complications develop, or to establish a diagnosis. There is a 25 percent incidence of recurrence if partial resection is performed.[1] Radiation can induce malignant transformation and hence it is not recommended.[9] Malignant degeneration can occur spontaneously in 0.5 percent of cases with a median total of 12.5 years from the time of diagnosis.[1] Patients with Fibrodysplasia are 400 times more likely to exhibit malignant bone tumors than are members of the general population.[4]

CONCLUSION

Fibrodysplasia may present in the elderly population and should be considered in the differential diagnosis of lytic skull lesions.

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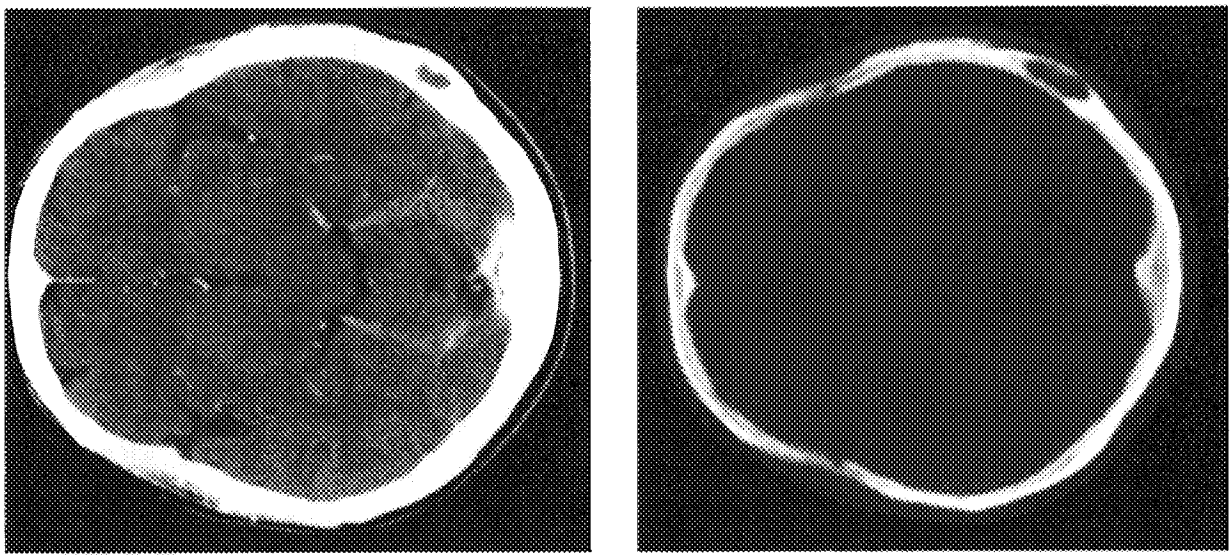
Figure 2a and 2b. CT scan showing lytic skull lesion in left posterior temporal area



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