Histiocytosis

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Lichtenstein (1953) suggested the term histiocytosis to embrace three syndromes which show the common origin of inflammatory reticuloendotheliosis. Each has distinctive clinico-pathologic features as a different manifestation of the same basic disorder.

Histiocytosis may range from an isolated slowly growing lesion particularly in the medullary cavity of bone to an aggressive widely disseminated disease with fatal outcome. Hence separate descriptive names were given:

(1) *Eosinophilic granuloma:* for bone lesions only, in children and young adults.

(2) *Hand-Schüller-Christian disease:* Chronic slowly advancing form in children and young adults. Manifestations resulting chiefly from bone involvement.

(3) *Letterer-Siwe disease:* Acute illness of infancy and early childhood, with a pattern of deeper more rapid visceral spread often involving bone as well.

A short review will be given on eosinophilic granuloma which is the topic of this paper.

Eosinophilic granuloma is a disease of children and young adults characterized by localized single or multiple skeletal lesions. Any bone in the body may be involved, but there appears to be a choice for the skull, extremities, pelvis, ribs and spine. The skull lesions have an affinity to the temporal bone, specially its squamous and mastoid parts and to the frontal bone. Less commonly the occipital bone, skull base or mandible are involved. Little or no systemic manifestations of the disease may be present.

Locally the disease manifests itself by painful and tender swellings on the affected bone. When the temporal bone is involved, there may be swelling over the mastoid region, otorrhea, granulations in the external ear canal, deafness, sometimes facial paralysis which may suggest tuberculous mastoiditis.

The frequency of associated ear discharge in cases of eosinophilic granuloma brings most of these cases first to the care of the otologist.

Deafness in these cases is due either to interference with the middle ear mechanism, or due to involvement of the inner ear.

In inner ear involvement the infiltration involves the otic capsule and destroys the endosteum (Lopez-Rios *et al.*, 1968). The slow invasion of the inner ear by cells of the disease explains the absence of vestibular manifestations.

Microscopically, the yellowish grey swellings contain numerous large histiocytes with a high eosinophil population (Fig. 1).

The pathologic stages have been described by Engelbreth-Holm *et al.* (1944) as follows:

(1) *Proliferative phase:* with histiocytes and eosinophils.

(2) *Granulomatous phase:* with increased blood vessels, reticular cells, giant cells and lipid phagocytosis.

(3) *Xanthomatosis stage:* with isolated foam cells.

(4) *Healing stage:* Fibrosis.

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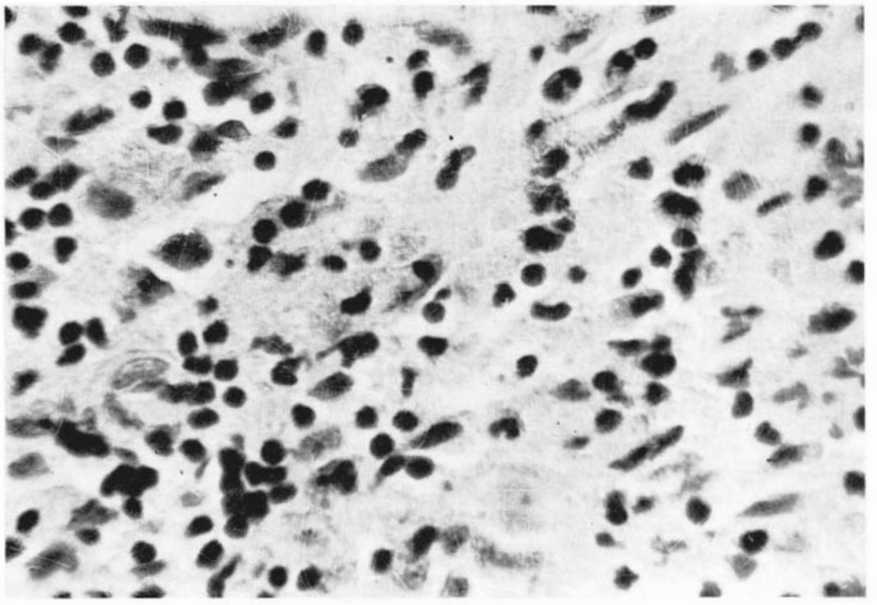


FIG. 1.

Prognosis of eosinophilic granuloma is guarded but favourable, as most patients recover. It is unfavourable in cases with multiple lesions and when there is visceral involvement.

As regards treatment, histiocytosis has no specific treatment. The following lines summarize the possible measures of treatment.

(1) *Surgery:* For localized lesions in bone only, currettage usually results in induction of healing. In mastoid lesions complicated with infection, mastoidectomy is indicated.

(2) *Radiotherapy beneficial in:*

(a) Cases with solitary lesions of bone.

(b) Produces rapid suppression of lesions in weight-bearing bones where pathologic fractures may result.

(c) In pituitary-hypothalamic lesions which causes diabetes insipidus.

(d) In troublesome cutaneous lesions and in adenopathy.

(3) *Drugs:*

(a) *Alkylating agents:* (as nitrogen mustard, chlorambucil, cytoxan) produce definite suppression of the disease. In children with bone lesions and moderate visceral disease, these drugs slow down the clinical progress and may be considered as life saving.

(b) *Vinblastine:* as an alternative to alkylating agents, is of definite value.

(c) *Corticosteroids:* Give additional symptomatic assistance to the patients with disseminated disease specially in regard to systemic symptoms.

(4) *General supportive care:* as blood transfusions, antibiotic therapy, orthopaedic care, and speech rehabilitation in deaf children.

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Clinical records

Report of five cases

Case No. 1

P.K., a 2-years-old female came with the complaint of discharging left ear, painful swelling of the forehead, and protrusion of the right eye.

Examination revealed a serous discharge from the left ear, with pale soft granulations in the meatus, and a retro-auricular swelling over the left mastoid region. There was also proptosis of the right eye down and out, and a definite firm swelling could be felt in the medial part of the roof of the orbit. Mobility of the eye was normal, and the fundus was also normal. The forehead was the seat of soft tender swellings. No nasal or nasopharyngeal disease. No lymph-adenopathy or hepatosplenomegaly.

Radiography showed bony defects in the frontal bone extending to the orbital plates specially on the right side (Fig. 2). Lateral view showed well delineated bony defects between the bony tables of the frontal bone. There was also well defined destruction in the left mastoid cells (Fig. 3).

Exploration of the left mastoid revealed bony destruction with the presence of soft yellowish granulations, and exposed sigmoid sinus dura. The posterior bony meatal bone was eaten up, and the middle ear full with the same type of

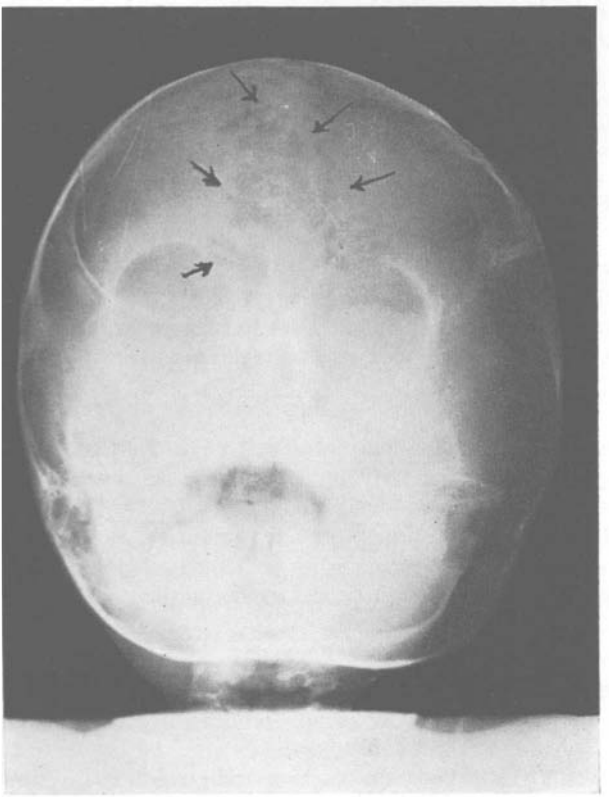


FIG. 2.

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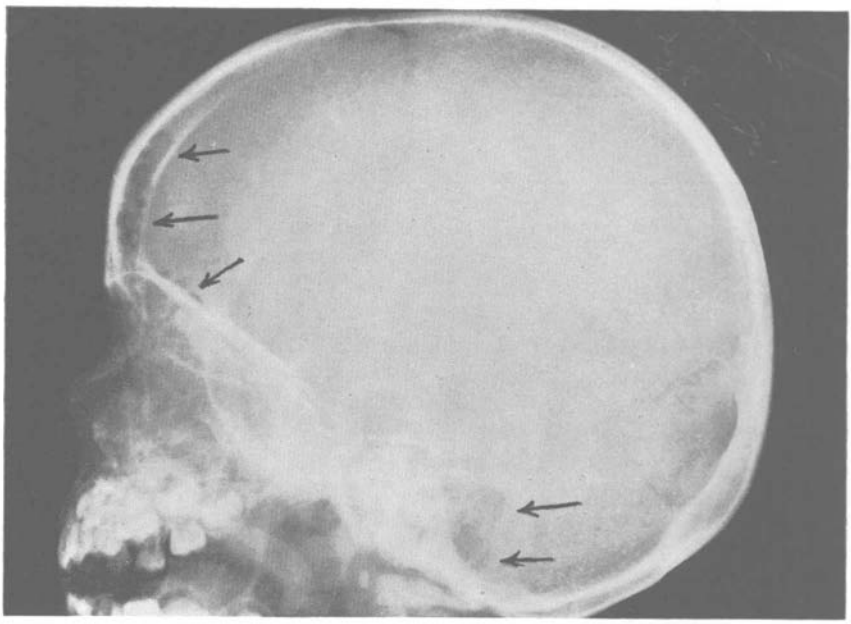


FIG. 3.

granulations. No trace of malleus or incus, but the stapes was found covered with the same granulations. The facial bony canal was destroyed, and the nerve exposed. As much granulation as possible was removed, and a radical mastoid completed. At the same time biopsy was taken from one of the frontal swellings.

Histopathology revealed a picture of eosinophilic granuloma.

Postoperative deep irradiation was given; the mastoid wound healed satisfactorily and the frontal swellings and proptosis disappeared. The girl was put on prednisolone for almost a year. The girl reports for follow up every month. For the last three years no evidence of recurrence has been detected so far.

Case No. 2

O.A.B., a 5-years-old boy presented with multiple swellings in the right temporal region, these were soft in consistency with well defined margins. There were also multiple firm swellings in the chest wall over the line of the ribs. The child had a slight limp, the general condition was fair with a low grade fever. No lymphadenopathy, skin rashes or hepatosplenomegaly. Blood picture showed no immature cells, but there was an increase in eosinophils (4 per cent).

Radiography

In the skull there were well defined bony defects in the lower lateral part of the frontal bone, in the greater wing of sphenoid and in the squamous temporal (Fig. 4).

Chest showed areas of bony destruction and erosion in the ribs (Fig. 5).

Pelvis showed large osteolytic punched out areas in both iliac regions (Fig. 6).

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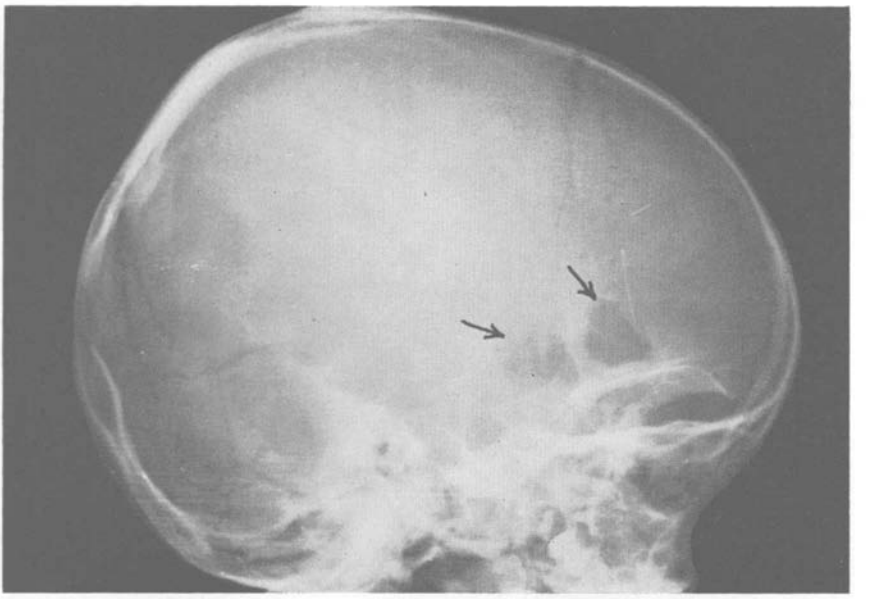


FIG. 4.

Biopsy from the temporal swelling revealed the picture of eosinophilic granuloma.

Deep X-ray treatment was given to all the sites of the lesions and at the same time prednisolone was given. Radiotherapy course was repeated twice in three years time as new lesions appeared. The patient is still on corticosteroids. X-rays show that while some of the original lesions remain, there is definite evidence of good healing.

Case No. 3

A.S., a 6-years-old boy presented with:

painful swelling in relation to the right ear for few days;
discharge and deafness in both ears for few months;
painful swelling of the top of the head.

Examination revealed a typical right mastoid abscess. Both ears were full of purulent discharge and pale soft granulations. There was almost total deafness as shown by different hearing tests. The head swelling was soft with well defined margins in the upper part of the parietal region.

Radiography showed extensive multiple areas of bone destruction in both mastoids with delineation of the bony labyrinth. There were multiple areas of destruction in the parietal bones (Figs. 7, 8, 9).

Right cortical mastoidectomy was done, the cavity cleaned of all granulations, and wound closed with drainage.

Histopathology revealed a picture of eosinophilic granuloma. Unfortunately treatment could not be continued as we lost contact with the patient.

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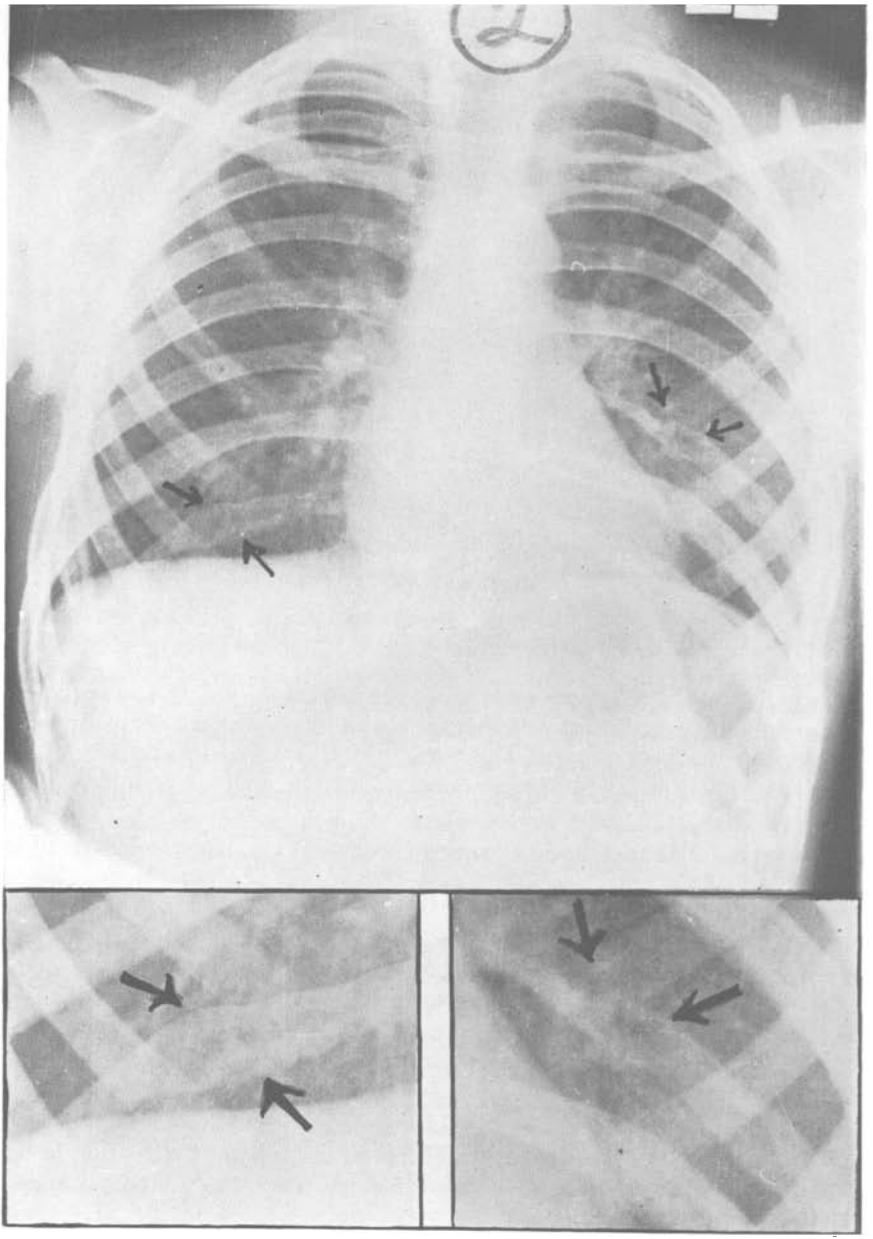


Fig. 5.

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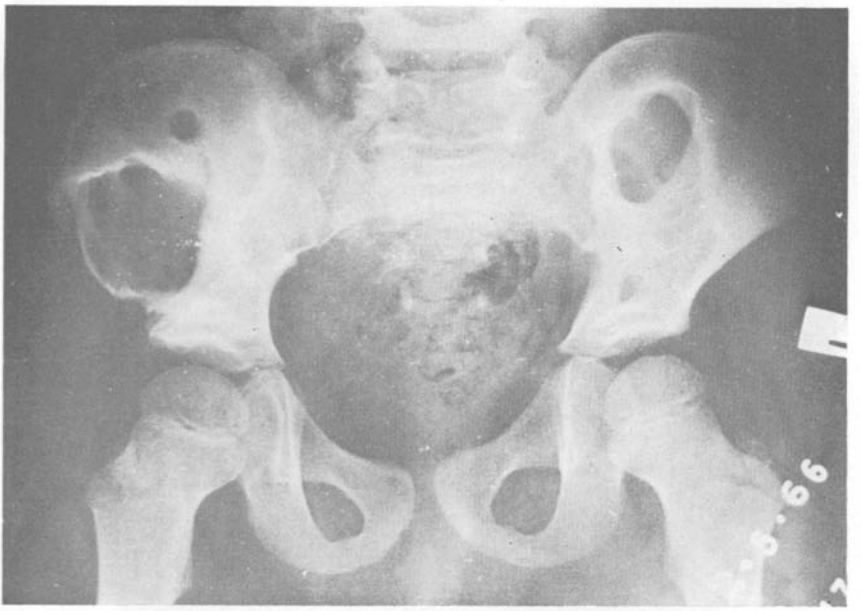


FIG. 6.

Case No. 4

A.S., 4-year-old boy presented with a swelling in relation to left ear, the left temple, cheek and left eyelid, and foetid purulent left ear discharge. There was also neck stiffness.

Examination revealed a typical left zygomatic mastoid abscess, with foetid purulent ear discharge. The ear was full of greyish haemorrhagic soft granulations. Neurological examination revealed no abnormal signs.

Radiography showed sharply defined bony defects in the left mastoid region and in the squamous temporal. There was also a well punched out osteolytic defect in the occipital bone. The body of the second cervical vertebra was involved with a lytic infiltration (Fig. 10).

Extended left mastoidectomy was done with currettage of all granulations. Tegmen and sinus plates were eaten up, and the dura of the middle and posterior cranial fossae exposed. There was no sign of any of the middle ear contents. Cavity was cleaned as far as possible, and the wound closed with drainage.

Biopsy revealed a picture of eosinophilic granuloma.

Postoperative irradiation was given and the wound healed uneventfully. The neck stiffness disappeared in a few days. He has been under prednisolone treatment for the last 7 months with no signs of progress or recurrence and with very good radiological healing of the skull and vertebral lesions.

Case No. 5

H.M., a $2\frac{1}{2}$-year-old male, came with left sided resistant otorrhoea that responded to no conservative treatment. He disappeared for some weeks to come back with a typical picture of left mastoid abscess.

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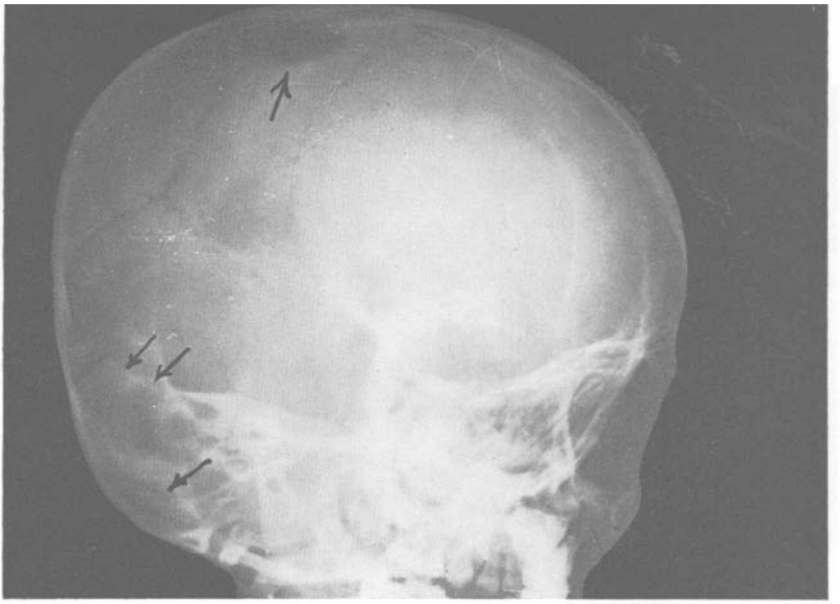


FIG. 7.

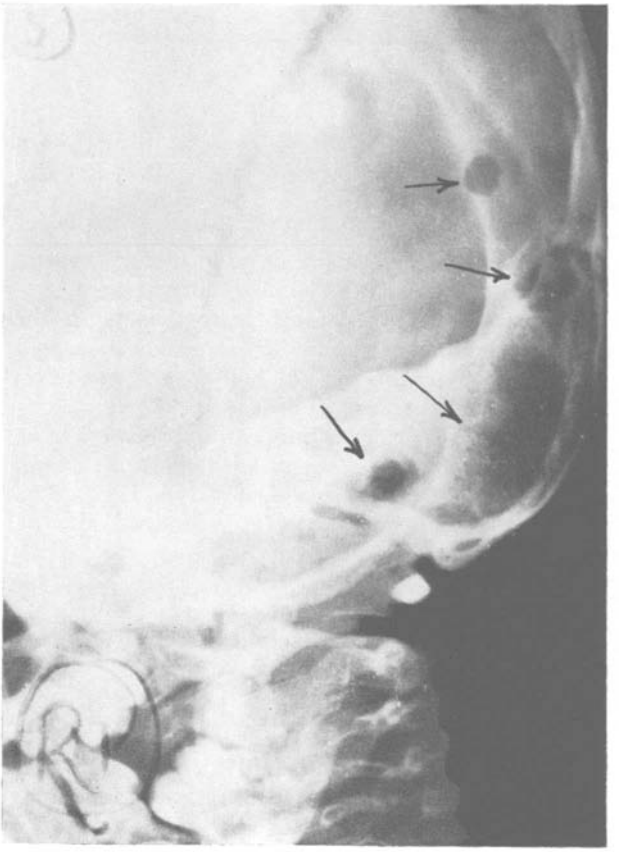


FIG. 8.

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FIG. 9.

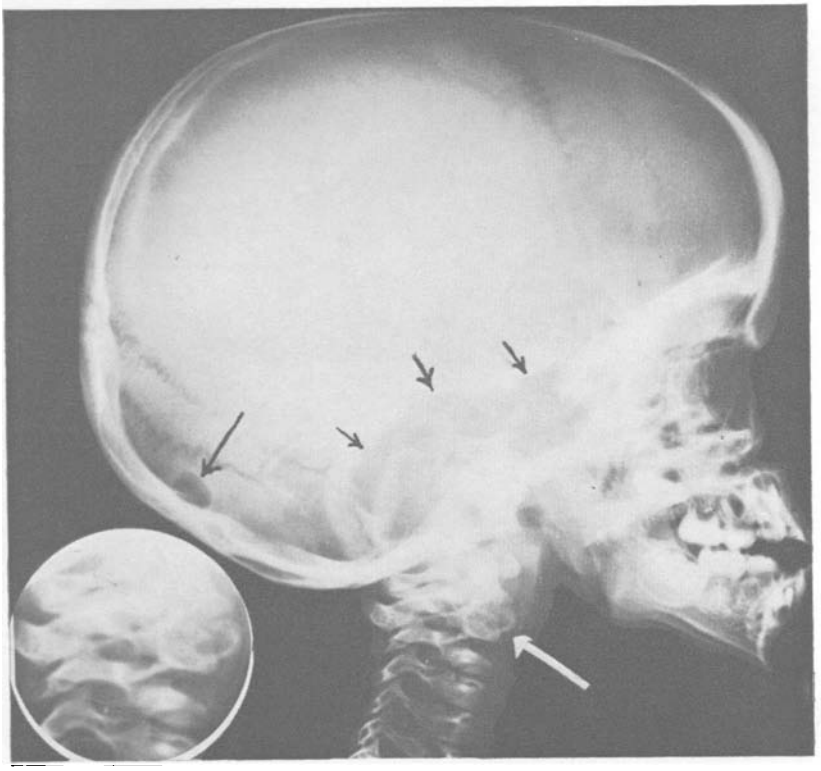


FIG. 10.

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Radiography revealed destruction of the left mastoid with well delineated bony defects in the mastoid and squamous temporal (Fig. 11).

Cortical mastoidectomy was done with removal of all the granulations. Biopsy revealed a picture of eosinophilic granuloma, therefore a radiologic survey of the skeleton was carried out. This revealed small area of infiltration in the right iliac bone of the pelvis (Fig. 12). Radiotherapy and corticosteroids were given. The wound healed satisfactorily, and the patient is still under observation.

Summary and conclusion

Histiocytosis is discussed in short with special reference to eosinophilic granuloma. The condition is further illustrated by the five recorded cases, all are children, the ages varying from 2 to 6 years, four males and one female.

Four of them were treated and followed up. They responded satisfactorily to deep irradiation and prednisolone treatment after surgical operation.

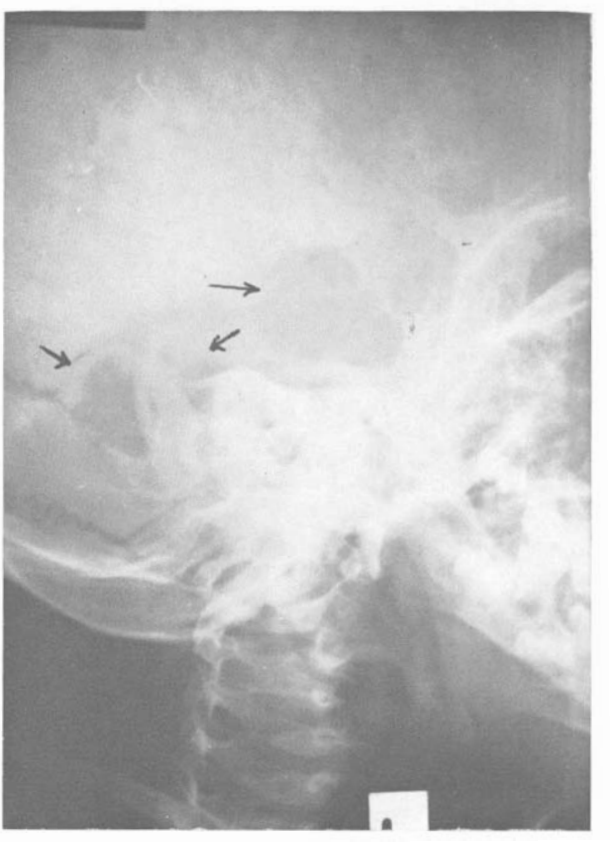


FIG. 11.

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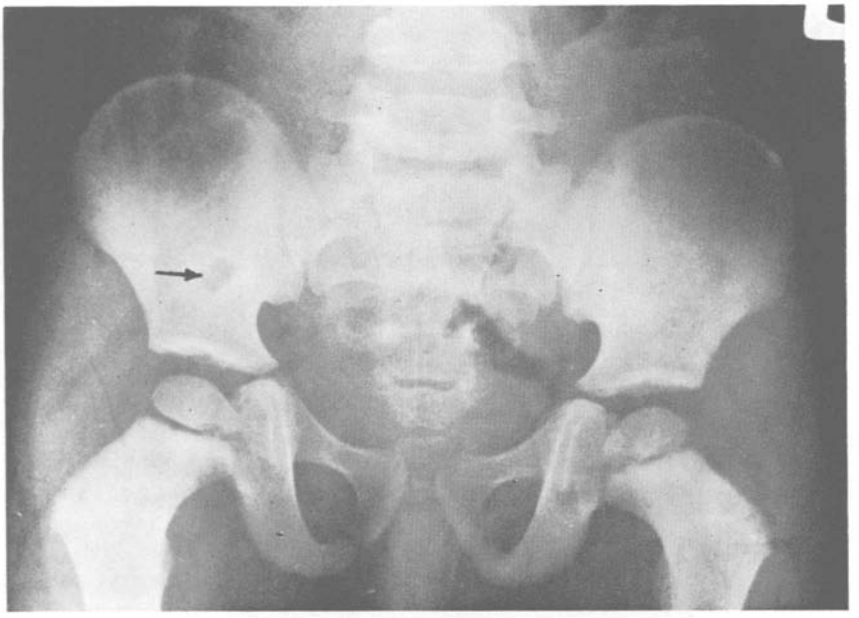


FIG. 12.

REFERENCES

AVERY, M. E., MCAFEE, J. G., and GUILD, H. G. (1957) *American Journal of Medicine*, **22,** 636.

CROCKER, A. C. (1968) *Current Pediatric Therapy*, Philadelphia, p. 488.

ENGELBRETH-HOLM, J., TEILUM, G., and CHRISTENSEN, E. (1944), *Acta Medica Scandinavica*, **118,** 292.

GREEN, W. T., and FARBER, S. (1942) *Journal of Bone and Joint Surgery*, **24,** 499.

KELEMEN, G. (1960) *Laryngoscope*, **70,** 1284.

LICHTENSTEIN, L. (1953) *Archives of Pathology*, **56,** 84.

LOPEZ-RIOS, G., BENITEZ, J. T., and VIVAR, G. (1968) *Annals of Otology, Rhinology and Laryngology*, **77,** 1171.

NELSON, W. E. (1969) *Text Book of Pediatrics*, 9th Edition. Philadelphia, p. 1479.

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