EOSINOPHILIC GRANULOMA OF THE TEMPORAL BONE WITH CUTANEOUS MANIFESTATIONS
(Case report)

* V. P. SOOD, † P. K. KAKAR AND ‡ I. S. GUPTA

Summary

A case of eosinophilic granuloma of temporal bone with cutaneous manifestations is presented. Eosinophilic granuloma is of special interest to the otologist because of its presentation in the temporal bone. The cutaneous involvement in this case is a rare event.

Treatment is excision and currettage of the bony lesions prognosis of this condition is guarded.

Eosinophilic granuloma is one of the reticuloendothelial granulomas affecting children and adults. It is of special interest to the otologist because of its presentation in the temporal bone. It belongs to a group of diseases described by Lichtenstein[1] under the name of Histiocytosis X; which includes eosinophilic granuloma, Hand-schuller christian disease and Litterer-Siwe disease. They differ clinically but have same basic process in form of proliferation of the reticuloendotheleal cells.

Eosinophilic granuloma is comparatively benign, usually localized to the skeleton and has been reported to occur in various bones, Mc-Gavron and Spady[2]. The lesion is frequently diagnosed in a skiagram because of its insidious onset. It involves the skeleton either in form of isolated lesions or multiple lesions. Besides affecting skeleton, it rarely involves skin, thyroid, pituitary, prostate, lungs and intestine, Samy[3]. In the skull it appears as a localised painful and tender swelling. Early small swellings are firm while larger ones may become soft in their central part and even fluctuant with ill defined edges. Cutaneous lesions may appear in form of papular eruptions, Curtis and Cawley[4]. If pituitary is involved patient gets diabetes insipidus.

Prognosis is good in single bony lesions.

* Assistant Professor of Otolaryngology, All India Institute of Medical Sciences, New Delhi.
† Professor and Head of the Department of Otolaryngology, Maulana Azad Medical College and Associated Irwin and G. B. Pant Hospitals, New Delhi.
‡ ENT Specialist, Holy Family Hospital, New Delhi.
Received for Publication on 12—2—1973

Case Report

Seven years old boy presented with the complaint of intermittent pain right ear for the last 10 months with gradually increasing swelling of the right postaural area for the last 3 months. There was history of skin eruptions on the chest, abdomen and back for the last 2 months. Patient complained of occasional slight mucopurulent discharge from the right ear.

Patient denied history of any ear discharge prior to this episode. There was no history of any urinary complaints.

On examination: well built fairly nourished with no lymphnode enlargement anywhere in the body. Locally, there was a 2 x 1 inch soft tender swelling behind right pinna extending on to the zygoma. There was oedema surrounding this swelling and occluding the retroauricular sulcus. The ear canal was found occluded due to sagging of posterior meatal wall. There was slight mucopurulent discharge coming out of the ear canal. Tympanic membrane was not visible. Left ear was normal.

Nothing significant noticed in nose and throat. Facial nerve was found intact.

There were extensive maculo-papular eruptions on the whole of the chest, abdomen and back. Nothing abnormal found on systemic examination.

Incision and drainage of the abscess was tried but no pus came out.

Investigations

Routine blood tests were normal. Urine normal. Audiogram: conductive loss of 40--50 dB in the right ear. X-ray mastoid: revealed punched out irregular defect around the mastoid region Fig. 1. Whole of the skeleton was radiographed but no other lesion was noticed in the bony skeleton.

Operation

Radical mastoidectomy was done giving post-aural incision. Immediately under the periostium, yellowish fatty necrotic tissue like mass was seen extending into the deeper planes of the surrounding soft tissues. Mastoid cortex was found destroyed. The yellowish soft mass was involving whole of the mastoid, destroying posterior meatal wall and extending into the zygomatic cells. The tip of the mastoid process, sinus and dural plates were found destroyed. This mass was extending into the aditus, middle ear and destroying a part of tympanic membrane. Incus was found missing. Facial nerve was spared. Whole of the mass was curretted out and sent for histopathology, the report was eosinophilic granuloma Fig. 2. Skin biopsy report first time revealed only chronic inflammatory tissue. Repeat biopsy of the skin lesion was reported Histio cytosis Figs. 3 and 4.

Fig. 1

X-ray mastoid (right) revealing punched out irregular defect around the mastoid region.

Discussion

Eosinophilic granuloma is considered to be a chronic inflammatory granulomatous lesion of unknown etiology probably viral in origin affecting chiefly the reticulo-endothelial system.

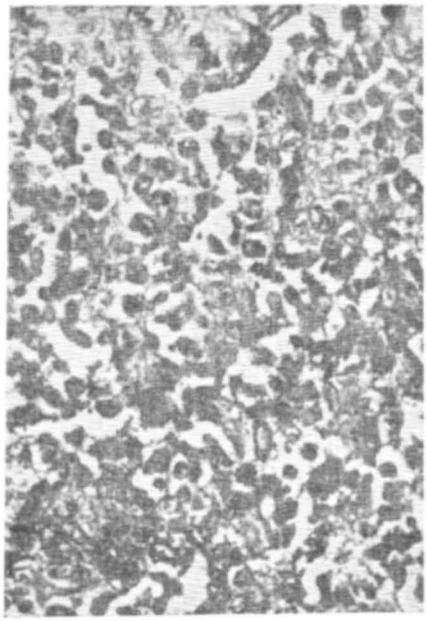

Fig. 2
Section from the mastoid region showing large number of histiocytes and many eosinophils, the eosinophils appearing as dark dots in the microphotographs E X 450

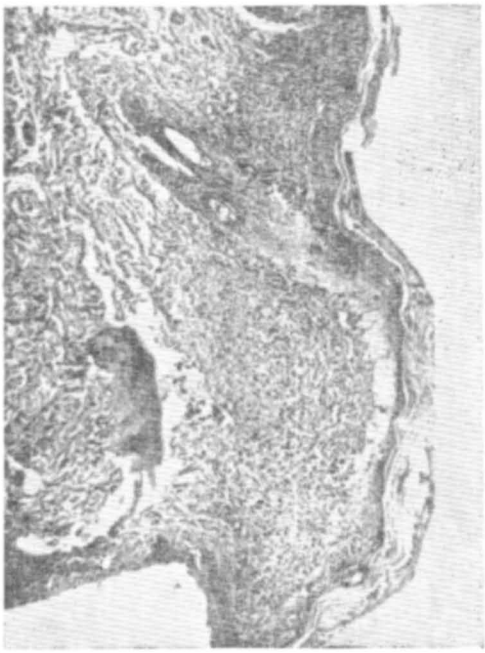

Fig. 3
Section of the skin, showing circumscribed involvement of upper dermis by the histiocytic process. H and E x 100

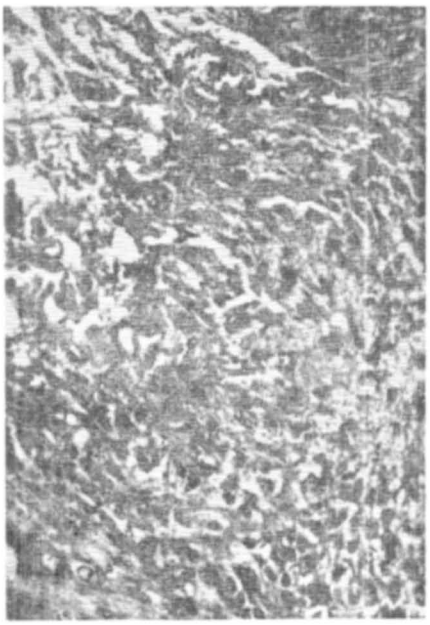

Fig. 4
Higher magnification of Fig. 3 reveals the details of histiocytes along with a fair number of eosinophils scattered throughout. H and E x 450

Lichtenstein[1] (loc. cit) grouped it under the name of Histiocytosis X and believe that eosinophilic granuloma, Hand-Schuller christian and Litterer-siwe diseases are interrelated manifestations of a single nosologic entity.

Mc Gavron and Spady[2] (loc. cit) are not impressed by the concept of reticulo-endotheliosis in which eosinophilic granuloma is considered to be the localized form and Hand-schuller christian and Litterer-siwe diseases are thought to be the disseminated types.

The transformation of eosinophilic granuloma to Handschuller christian and Litterer-siwe diseases must be if it does occur at all, very uncommon. They did not observe this transformation in 28 cases. They feel that infective etiology cannot be ruled out until meticulous microbiological investigations are carried out.

Samy[3] described this group as a disease involving lipoid metabolism chiefly due to the accumulation of cholesterol in these conditions.

The case under discussion was suspected clinically as a case of eosinophilic granuloma which was confirmed on radiological and histopathological examination. The conductive deafness present in this case was due to the involvement of middle ear with eosinophilic granuloma.

Histiocyte is the main cell of the cellular infiltrate in eosinophilic granuloma. The histiocyte nodule may contain plasma cells, lymphocytes and eosinophils. In later stages fibrosis may appear. The scarring and foam cells present in Handschuller christian disease are regarded as late manifestations of the eosinophilic-granuloma.

An eosinophilia of 6–10% may or may not be present. The blood cholesterol, serum phosphate, serum calcium and serum proteins are normal, Dawes[5].

The cutaneous lesions in the present case were in form of maculo-papular eruptions on the chest, abdomen and back and were proved histologically as histiocytic lesion. Manifestation of cutaneous lesions in eosinophilic granuloma is a very rare event.

Curtis and Cawley[4] and Mecreary[6] each have reported a case of eosinophilic granuloma with cutaneous manifestations.

Treatment of this condition has included currettage and small doses of radiation. Prognosis is guarded but it is good in single bony lesions.

Acknowledgement

The authors wish to thank Director-Principal, Maulana Azad Medical College and Associated Irwin & Pant Hospitals for permission to publish this case.

REFERENCES

1. Lichtenstein L: Histiocytosis-Integration of Eosinophilic granuloma of bone "Litterer-siwe disease and schullar christian disease" as related manifestations of a single nosologic entity, AMA Arch Path 56 : 84, 1953.
2. Mc Gavron MH and Spady HA: Eosinophilic granuloma of bone (study of 28 cases, J Bone Joint Surg, 42A : 979, 1960.
3. Samy LL: Eosinophilic granuloma with a report of seven cases, J Laryng, 79 : 42, 1965.
4. Curtis AC and Cawley EP: Eosinophilic granuloma of bone with cutaneous manifestations. Report of a case. Arch Derm and Syph 55 : 810, 1947.
5. Dawes JDK: Eosinophilic granuloma (Eosinophilic granuloma, Litterer-siwe disease and Handschuller christian disease) J Laryng, 68 : 575, 1954.
6. Mecreary JH : Eosinophilic granuloma with simultaneous involvement of skin and bone, Arch Derm Syph, 58 : 372, 1948.