CASE PRESENTATION

Eosinophilic Granuloma in E.N.T.

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INTRODUCTION

HISTIOCYTOSIS (H-X) is a term which embraces three different forms of the same disease i.e. eosinophilic granuloma, Hand-Schuller-Christan's disease (H.S.C.), Litterer Siwe disease (L.S.) Lichtenstein 1953).

Historically Histiocytosis X has been considered as three separate disease.

1. Hand-Schuller Christan's disease usually affects children in a chronic form with diabetis insipidus, lytic lesions of bone and exophthalmos.
2. Litterer Siwe disease — a rapidly fatal histiocytic infilration of many organs, principally affecting infants and young children.
3. Eosinophilic granuloma is confined to bone with solitary or multiple lytic lesions. Lung involvement is not an uncommon occurrence during the 3rd and 4th decades of life.

Eosinophilic granuloma, Hand-Schuller-Christan's disease, Litterer Siwe disease may be different forms of the same disease as put forward by Wallgren (1940) and Farber (1941).

The concept of these three conditions belongs to the same disease is strengthened by the findings of intracytoplasmic structures in abnormal histiocytes in all three conditions, thus suggesting a single aetiology. This has been found by electron microscopic studies which showed rod-like structure in the cytoplasm of abnormal histiocytes. The structure which appears to have constant form invariably contain central filaments (Booth and Thomas 1970). They have never been found in the nucleus or outside the histiocytes. This finding suggests a viral aetiology, yet to be proved.

Eosinophilic granuloma was originally described by Finzi, (1929) later by Otani and Ehrlich (1940) and Lichtenstein and Jaffie (1940) as a tumour — like destructive swelling of bone, usually solitary but occasionally multiple. The lesion occurs in skull, nose, sinuses, ribs, pelvis, scapula, clavicle and long bones, but never in the bones of the hand and feet (Mallory, 1942, Toss 1966). This varient usually affects older children and adults, but can also be seen in infants and the elderly. The lesion is usually osteolytic and circumscribed, but a diffuse sclerotic lesion has been reported (Ochsner, 1966).

Four Phases have been described in the early stages of eosinophilic granuloma, but usually they merge into each other.

The pathological stages described by Engelbreth Holme et al, (1944) are :-

1. Proliferative phase with histiocytes and eosinophils.
2. Granulomatous phase with increased blood vessels, reticulum cells, gaint cells and lipid phagocytosis.
3. Xanthomatous phase with isolated foam cells.
4. Healing stage with fibrosis.

Eosinophilic granuloma is infrequently seen in E.N.T. practice. It can involve nose, lips, paranasal sinuses, temporal bones and larynx (Booth and Thomas, 1970).

CASE REPORT

Case No. 1
M.H.N.M. — male, aged 26 months.

The above child attended the E.N.T. clinic on 24.8.1972 with a history of pain in the ear for one week. There was no history of ear discharge and no other specific complaints. General examination revealed a slightly febrile child in good general condition. E.N.T. examination revealed a tender

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swelling over the left mastoid tip. The left T.M. appeared dull. The patient was admitted with a provisional diagnosis of acute mastoiditis.

X-ray of the mastoids showed large antra on both sides. Blood examination showed that the child was anaemic (Hb 7.2. gm%).

Within two days a fluctuant postaural swelling developed. The T.M. could not be seen as the posterior wall of the left external autitory canal was oedematous.

On 27.8.1972 the left ear was examined under anaesthesia and incision and drainage of the abscess carried out. E.U.A. of the left ear showed boggy T.M. The shadow of malleus was not identified. Incision allowed drainage of a subperiosteal yellowish jelly-like tissue. A massive bony defect involving the lateral aspect of mastoid was seen. Two large defects were seen exposing the dura over the tegmen tympani and tegmen antra and also a large exposure of the sigmoid sinus. All the granulation tissue over the dura, additus and antra cleared. The facial bridge was removed and the ridge lowered. The malleus and incus removed and the granulation tissue around the stapes cleared though not completely. A large fistula was seen over lateral semicircular cannal. The seventh nerve was found exposed. Perifacial cells were involved. The cavity was packed with BIPP. All the granulation tissue was sent for histopathology and reported as showing the proliferative phase of eosinophilic granuloma. Post operatively radiotherapy was advised but the parents refused. So prednisolone 10 mg. bd. was given for one month and asked for regular follow-up.

The patient was seen on 3.7.1973 with history of blood stained otorrhoea. Examination revealed a friable mass filling the left cavity and external auditory canal. Admission was advised but refused. Later the patient was admitted on 26.4.1975 with the same complaints. Examination revealed the same ear findings and the child was deeply jaundiced with enlarged liver up to the umbilicus.

Patient was put on Vinblastin 1.4 mg daily (Wt. 15 Kg) for three weeks and prednisolone 2 mg/kg/day for four weeks. He was discharged but readmitted on 10.3.1975 with involvement of both ears by Eosinophilic Granuloma.

He developed meningitis secondary to CSF leakage from both ears. No more surgical intervantion could be offered due to his poor general condition. He was discharged and readmitted on 24.5.1976 with breathlessness, ascitis etc., which did not respond to treatment and the child died on 2.6.1976.

Other Investigations

Hb - 7.2 mg%, Wbc - 14, 800/cu mm, N 71%, E 1%, M 2%, *L 26%.*

Serum bilirubin, 10 mg/ 100 ml.

Glutamic Pyruvic transaminase 115 units/ml.

Ears swab — Negative when seen with bilateral involvement.

Case No. 2

M.M.R. — male, aged 16 years.

The above patient was seen on 25.5.1977 with the history of :

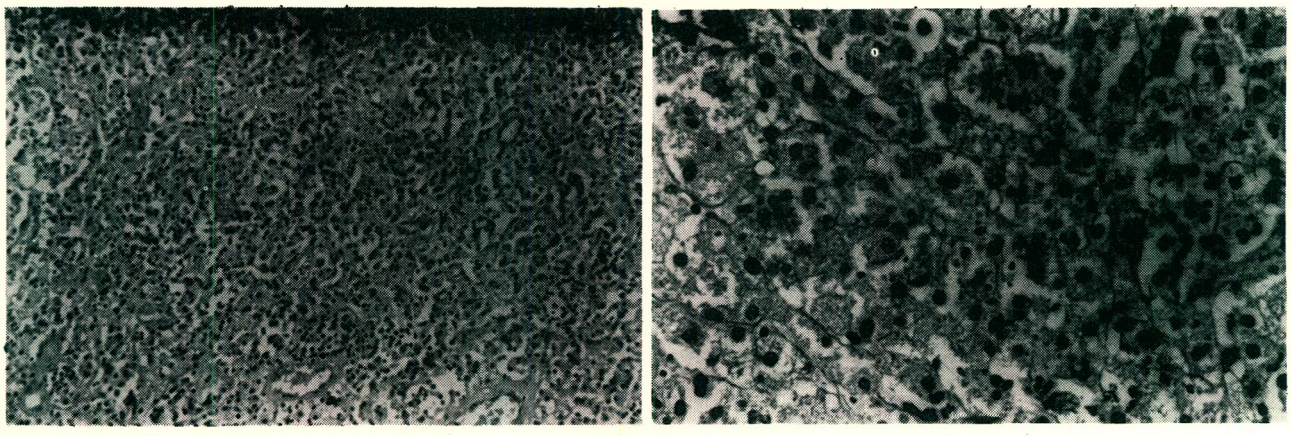
Bilateral Otorrhoea since childhood.

Blocked left ear — two months.

Left postarral swelling — two months.

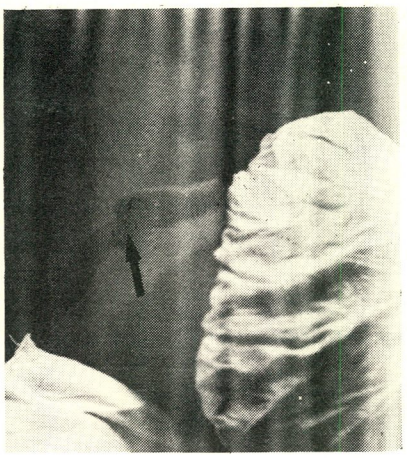
Left severe otalgia — two weeks and fungating mass in the left infra auricular region — one week.

The case started by the appearance of a left postaural swelling two months earlier which increased in



These are sheets of large reticulam cells with abundant granular cytoplasm and large central vesicular nuclei. These are admixed with variable numbers of neutrophils.

Fungating mass seen in the external meatus



Infra Auricular swelling seen below the left ear

size until it ruptured leaving a fungating mass associated with profuse left otorrhoea. Significant findings in the past history are childhood poliomyelitis and jaundice.

General examination revealed Kyphosis and Pigeon chest. E.N.T. examination revealed an active right CSOM and on the left ear there was swelling behind the pinna which was pushing the pinna forward. There was a fungating mass in the left infra-auricular region. The left external auditory canal was obstructed by granulation tissue which was protruding upto the concha. He was admitted with a provisional diagnosis of Eosinophilic Granuloma, Malligant Parotid Tumour, or Chronic Mastoiditis. X-ray showed sclerotic mastoids. Haemotological examination was within normal limits. Ear swab grew Staph aureus and Klebsilla.

He had an EUA of the left ear followed by radical mastoidectomy. EUA showed that the external ear canal was full of Granulation tissue which was removed and sent for histopathology. Granulation tissue was seen passing through a fistula from the external auditory canal leading to the parotid gland and fungating then and there. The mastoid bone exposed up to the tip. The antrum, additus and attic were filled with granulation tissue. All granulation tissue was removed and the lateral sinus and dura were exposed. The mastoid cavity was cleared upt to the tip. The short process of incus was covered with granulation tissue. The head of the malleus was missing; the rest of the malleus was removed and so was the incus. The facial bridge was removed and the ridge lowered. Meatoplasty was done and the mastoid cavity filled with BIPP. The biopsy was reported as showing eosinophilic granuloma. Regular follow up showed healing of the Cavity with no evidence of recurrence. Radiotherapy was thought unnecessary in this case.

Other Investigations

1. Skin test for allergy - nagative.
2. Skeletal survey - normal.
3. Blood examination showed eosinophils 4%.

DISCUSSION

Aetiology

The aetiology of histiocytosis X is still unknown and various aetiological factors are considered.

1. Bacterial
2. Mycotic
3. Fungal

Viral aetiology is based on electron miscoscopic findings of intra cytoplasmic tubular structure. But convincing evidence is still missing. Some authors believe it, as manifestion of hypersensitivity. Age incidence — Usually children under 15 years old are affected (Entriquez and Liberman).

Sex — Males are more frequently affected than females.

Predisposing Factor

Chronic Suppurative otitis media may predisposes to this condition.

Site of lesions in eosinophilic granuloma

Bone – Skull, nose, sinuses, pelvis, longbones. The lesion is usually lytic but rarely sclerotic. Eosinophilic granuloma is characterised by single or multiple skeletal lesions. Any bone can be involved but there appears to be a predilection for skull bone. The skull lesions have an affinity for the frontals and temporal bones especially squamous and mastoid. Less commonly occipital, skull-base and mandible are involved.

(Little or no systemic manifestitions are present). The lesion presents locally as a painful and tender swelling.

Once the temporal bone is involved there may be swelling over the mastoid region, otorrhoea, granulation tissue in the external ear, deafness and some-

times facial nerve palsy which may suggest a tubercular mastoiditis. The frequency of ear discharge brings the patients to the otologist. Deafness is due to involvement of the middle and inner ear. Seventh nerve and labyrinthine involvement have been described by Lapez Rios in 1968, Straka and Caparosa in 1972.

Skin:-

Papillomatous, Usually in children, Ulcerative in adults.

Oral lesion

Teeth. (Periodontal, Periapical) Jaws could be affected.

Cerebral and Cerebellar Dysfunction

Recently Braunstein et al reported cerebral and cerebellar dysfunction in four cases.

Endocrine dysfunction

Diabetics insipidus and thyroid dysfunction through hypothalamic axis involvement or not has yet to be established.

Macroscopic appearance

Mixture of cells including lipid laden macrophages, multinucleated giant cells, but predominently eosinophils (Muir).

INVESTIGATIONS OF HISTIOCYTOSIS X

1. Blood film which shows leucocytosis with relative neutrophilia, and mild Eosinophilia (3 to 10%), E.S.R. within normal or slightly elevated.

2. Sputum (for lung involvement) may show large numbers of Eosinophils or fat laiden histiocytes.

3. Skin test for allergy.

4. Full radiological survey including skull. Poly-phosphate bone scan might be valuable because early lesion can be picked up.

5. Biopsy of the lesion.

6. Recently Electron miscroscopic studies have proven to be valuable as mentioned earlier (Booth)

TREATMENT

Four types, either single or in combination.

Surgery

The disadvantage of surgical treatment is the limitation of surgical interferance due to the location of the lesion.

Steroids

The disadvantages of steroid therapy are obesity, cushing's syndrome, activate peptic ulcer and possible perforation, osteoporosis, euphoria etc.

Intralesional steroid has been tried but with very limited success (Regression).

Radiotherapy

The disadvantages are perichondritis, subglottic oedema, late stenosis, development of carcinoms 20 to 30 years later, radionecrosis. Dermal eosinophilic granuloma is not radiosensitive and should be exicsed. Recurrence after excision has been recorded.

Cytotoxic Chemotheraphy

The disadvantages are agranulocytosis, alopacia, bone narrow depression, skin reaction .....etc.

PROGNOSIS

Proper management of non-osseous disease (diabetis insipdus and skin) might be chronic but non fatal according to Leiberman. Prognosis is favourable in single lesions and unfavourable in multiple bony lesions and in visceral involvement.

SUMMARY

Two cases of eosinophilic granuloma have been reported. The aetiology, site of lesions, and disadvantages of different forms of treatment have been discussed.

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