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Radiology in Focus

Langerhans' cell histiocytosis: temporal bone involvement

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Abstract
In Langerhans' cell histiocytosis, radiological findings on temporal bone involvement show destructive bone lesions involving the mastoid, with the squamous part and middle ear less affected. Computed tomography (CT) is the preferred imaging modality for describing the extent of temporal bone involvement. CT also has an important role in monitoring disease activity and response to treatment. Bone scintigraphy seems to be less sensitive than radiography in detection of these lesions.

Key words: Histiocytosis; Langerhans' Cell; Temporal Bone; Tomography; X-Ray Computed; Scintigraphy; Bone

Langerhans' Cell Histiocytosis

Langerhans' cell histiocytosis (LCH) is the accepted term to describe the disease referred to as eosinophilic granuloma, histiocytosis X, Hand-Schüller-Christian disease, or Letterer-Siwe disease. LCH is principally a paediatric disease (incidence in the paediatric population of three cases per 1 000 000 children per year), characterized by the proliferation of histiocytes, many of which present the Langerhans' cell phenotype,accompanied by a proportion of multinucleated giant cells and eosinophils.[1] Accumulation of abnormal histiocytes has been described in various organs including the bone marrow, skin, larynx, lung, lymph nodes, thymus, liver, spleen, thyroid, parotid gland, gastrointestinal tract, kidney and central nervous system.[2] LCH often affects the head and neck region. Di Nardo and Wetmore,[3] in their series of 100 patients, estimated the incidence of head and neck complaints to exceed 80 per cent of cases (63 per cent on presentation of the disease and an additional 20 per cent during the course of the disease).

The skull is the most frequently involved head and neck site, affecting, in decreasing order of frequency, meatal skin, cervical nodes, temporal bone, maxilla and mandible.[4] Temporal bone involvement has been described in 13.5 to 61 per cent of patients with LCH.[5,6] The disease may present with otological symptoms but no other clinical findings in five to 25 per cent of patients. Temporal bone lesions manifest as mastoid swelling, middle ear polyps, otorrhoea resistant to medical treatment, and erosion of the posterior bony external auditory canal. Conductive hearing loss may occur owing to obstruction of the external auditory canal or granulation tissue in the middle ear. Involvement of the bony labyrinth is rare, with few cases being reported in the literature. The bony labyrinth appears to be resistant to erosion by granulation tissue.[7] Cranial nerve paralysis is also unusual, although paralysis of the VIIth and VIIIth cranial nerve has been described.[8] Where temporal bone is involved, approximately 30 per cent of affected patients demonstrate bilateral disease.[8,9] Delayed diagnosis and diagnostic errors are frequent because otological findings are often similar to other conditions such as acute mastoiditis, chronic otitis media, cholesteatoma or external otitis.

Imaging investigation

Computed tomography (CT) is the preferred imaging modality to describe the extent of temporal bone involvement. It is also a useful tool for monitoring disease activity and response to treatment.

CT shows bone destruction, which may be bilateral and symmetric. Associated lesions can be seen in the squamous part of the temporal bone. In some patients, the mastoid process is involved and destruction of the auditory ossicles, extension to the bony labyrinth and lesions of the petrous apex are observed.

The lesions have indistinct margins and the smaller structures of the bony labyrinth and auditory ossicle chain may display erosion. However, involvement of the auditory ossicles and internal ear is not as frequent as might be expected from the extensive bony damage usually seen (Figure 1(a) and (b)).

Some patients have an associated soft-tissue mass, which almost always appears homogeneous after administration of contrast media (Figure 2(a) and (b)).

Extradural extension may be observed adjacent to the temporal lobe as a soft-tissue mass. In these situations, magnetic resonance imaging (MRI) is more precise than CT in showing both the extent of the lesion and its relationship to adjacent structures such as the temporal lobe and cerebellopontine angle. The soft-tissue mass shows strong signal intensity (hyperintense) on T2-

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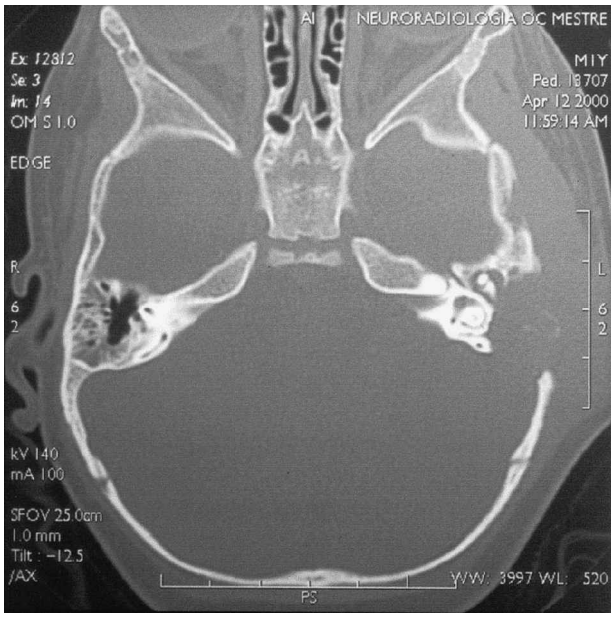
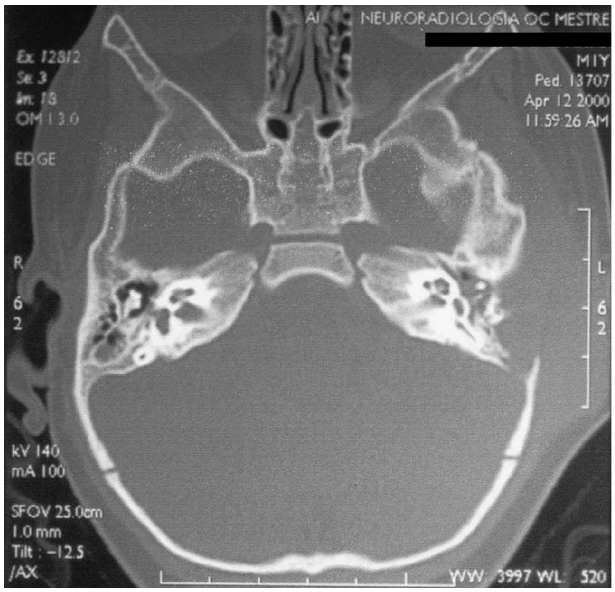


(a)



(b)

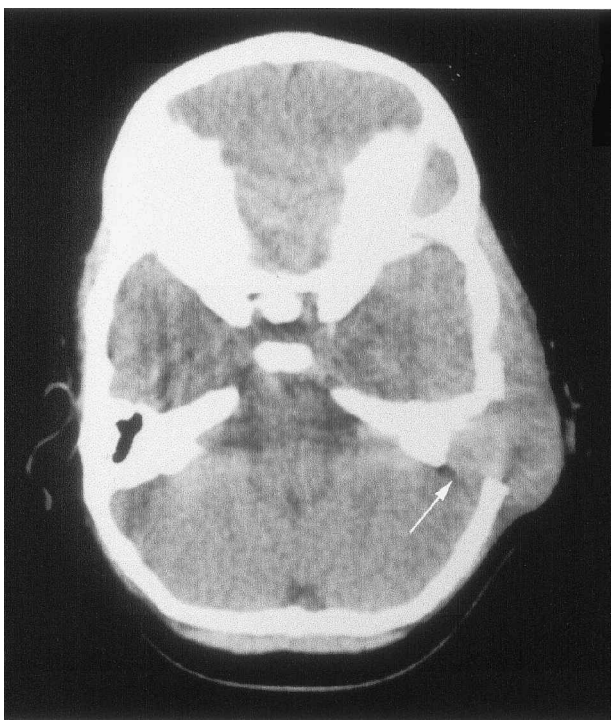
FIG. 1 (a) and (b)

(a) The left temporal bone shows a wide erosion area; although the middle ear is largely obliterated by soft tissue, the auditory ossicles are still intact (b).

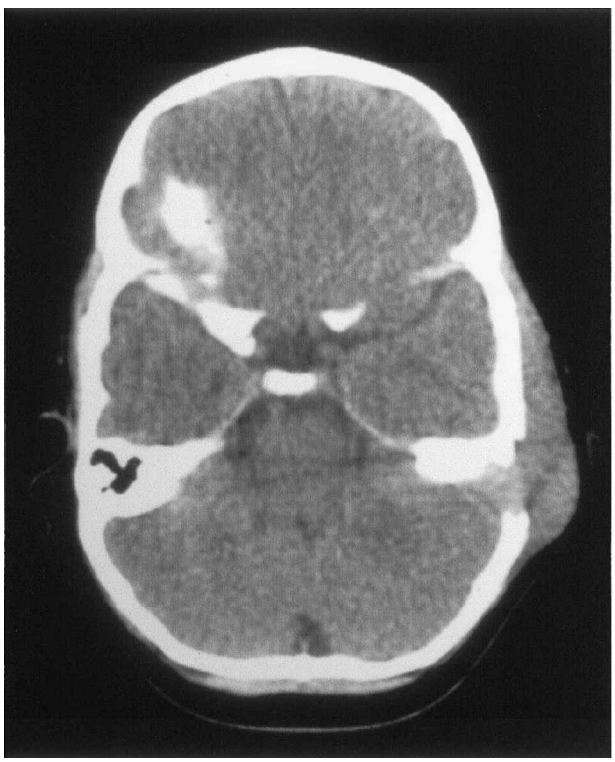
weighted images and variable signal intensity (isointense to hypointense) on T1-weighted images, often with peripheral oedema or inflammation around the lesion and with marked enhancement after administration of gadolinium.

CT delimits bony destruction of the petrous apex better than MRI. MRI more clearly delineates extension of the soft-tissue mass, owing to its multiplanar capacity and higher contrast resolution.

Skeletal radiology is generally regarded as the most accurate means of detecting LCH-related bone lesions. Bone scintigraphy appears to be less sensitive (Figure 3(a) and (b)) than radiography in detecting old lesions and a radiographic bone survey is preferred to bone scanning in identifying bone lesions in LCH patients. The sensitivity and specificity of radiographic survey for LCH detection were 100 per cent and 61 per cent, respectively, compared



(a)



(b)

FIG. 2 (a) and (b)

(a) soft tissue mass is clearly visible in the left temporal region reaching, without involvement, the posterior cranial fossa by erosion of temporal bone (b).

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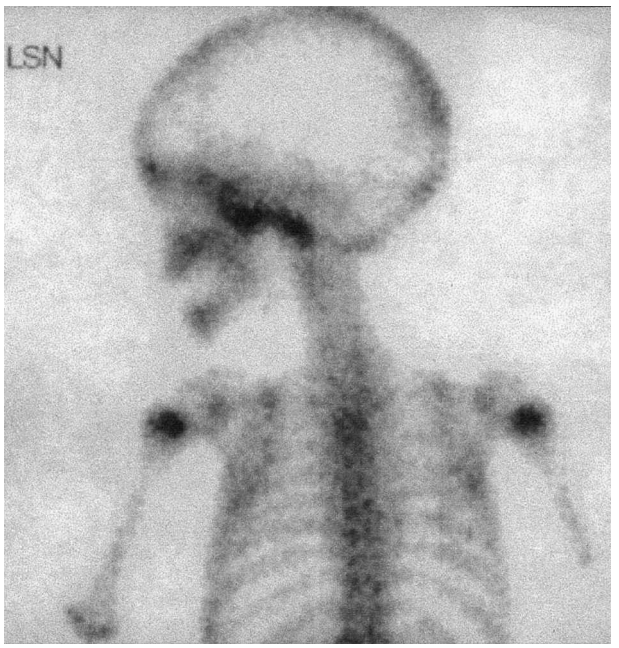
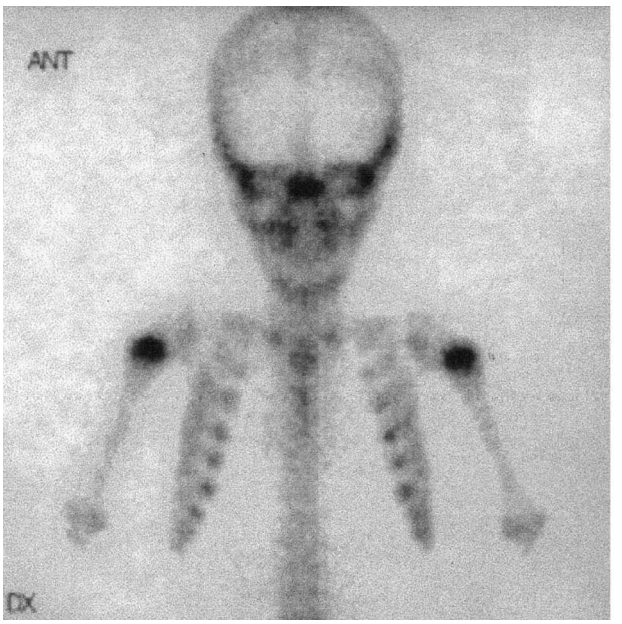


(a)



(b)

FIG. 3 (a) and (b)
Bone scintigraphy does not show significant uptake of radionuclides due to its lower sensitivity.

to 91 per cent and 55 per cent for bone scintigraphy. The accuracy of skeletal radiograph survey was 90 per cent compared to 82 per cent for bone scintigraphy,[10] but bone scintigraphy may reveal additional lesions, especially in complex bones. Furthermore, bone scintigraphy has been advocated to have a role in LCH diagnosis and staging.

The typical radiographical appearance of LCH is one of a shining lesion, sometimes well-circumscribed, often with sclerotic margins and a bevelled edge. On bone scintigraphy, LCH typically appears as a focus of avid tracer uptake or, alternatively, a circumscribed rim of increased radiotracer activity surrounding an area of low uptake. The radiological differential diagnosis includes mastoiditis, rhabdomyosarcoma and metastasis. Extensive destruction of bone and large soft-tissue mass is not usually seen in mastoiditis, but the other two conditions may have similar clinical and radiological findings. However, clinical presentation and radiological findings may diagnose Langerhans' cell histiocytosis, especially when bilateral involvement is present. A definite diagnosis is established on biopsy.

Conclusion

Langerhans' cell histiocytosis of the temporal bone presents as extensive bony destruction associated with a soft-tissue mass of variable size that usually enhances uniformly after administration of i.v. contrast material. Examination and follow-up with CT are the most accurate tools for evaluating the evolution of the radiological findings.

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