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Histiocytosis X of the petrous bone in the adult: MRI

A. Bonafé[1], H. Joomye[1], P. Jaeger[1], B. Fraysse[2], C. Manelfe[3]

[1] Service de Neuroradiologie, Hôpital Purpan, Toulouse, France
[2] Service ORL, Hôpital Purpan, Toulouse, France

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Abstract. Two cases of histiocytosis X in young adults with involvement of only the petrous bone are reported. In the first symptoms and signs consisted of a seventh nerve palsy of gradual onset over 3 years, and in the second, of pulsatile tinnitus associated with otorrhoea of clear fluid and impaired vestibular function. CT revealed large lytic lesions of the petrous bone. T1-weighted MRI before and after gadolinium confirmed the presence of a mixed tumour of the petrous bone with marked uptake of contrast medium. Spin echo images demonstrated the absence of extradural extension, and the patency of the arterial and venous network, therefore enabling differential diagnosis from extensive glomus jugulare tumours.

Key words: Histiocytosis X – Petrous bone – CT – MRI

Histiocytosis X is characterised by proliferation of cells having the same immunohistochemical and ultrastructural characteristics as Langerhans cells of the epidermis [1]. The three classical syndromes Hand-Schüller-Christian disease, Letterer-Siwe disease and eosinophil granuloma are now of only historical interest [2].

We report the computed tomographic (CT) and magnetic resonance imaging (MRI) findings in two cases involving the petrous bone.

Case reports

Case 1

A 16-year-old boy presented with a history of headache, progressive pulsatile tinnitus in the right ear, abundant otorrhoea of clear fluid, and intermittent vertigo. Otoscopy showed stenosis of the external auditory canal. The erythrocyte sedimentation rate was normal. Pure tore audiometry and brain stem evoked auditory responses (BEAR) showed mixed deafness on the right. Electronystagmography and electrocochleography did not reveal a right vestibular lesion, and electromyography of the facial nerve was normal.

High resolution CT of the right petrous bone showed a lytic, heterogeneous, enhancing mass, which extended to the labyrinth and to the third portion of the Fallopian canal. MRI, with T1-weighted axial and sagittal images showed the tumour, which was isointense with the adjacent brain parenchyma, to enhance after injection of contrast medium (Fig. 1). It contained several, small, rounded low signal cysts, with peripheral enhancement, which gave high signal on T2-weighted images.

Case 2

A 28-year-old man, with a history of otitis media recurrent for 3 years, presented with a left seventh nerve palsy, pulsatile tinnitus, deafness of the left ear, paroxysmal left-sided headaches and diplopia. Examination showed a large retroauricular swelling and palsies of the left sixth, seventh and tenth cranial nerves. Otoscopy revealed a stenosis of the external auditory canal, which contained numerous cutaneous debris. Pure tone audiometry and BEAR confirmed the left-sided deafness.

CT and MRI examinations revealed a lytic tumour of the left petrous bone. T1-weighted images (Fig. 2) showed numerous small low signal areas within the isointense mass. The latter enhanced ho-

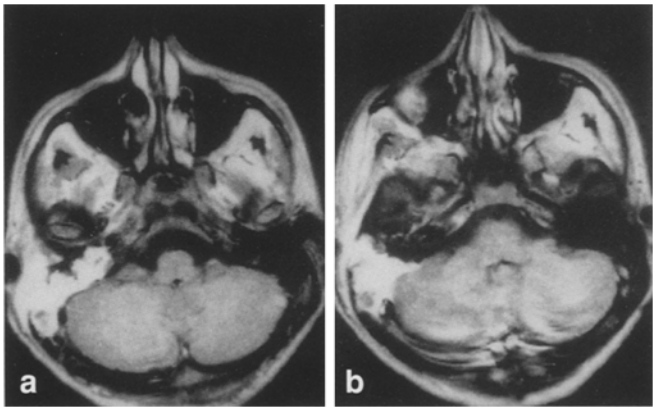
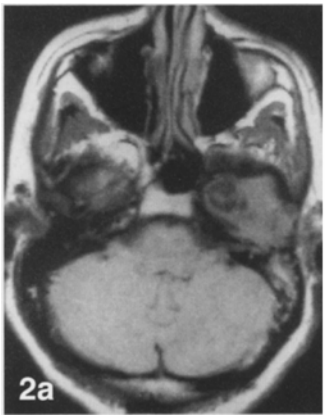
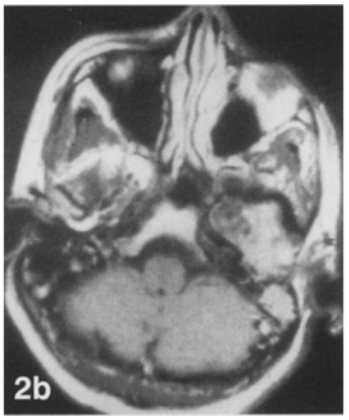
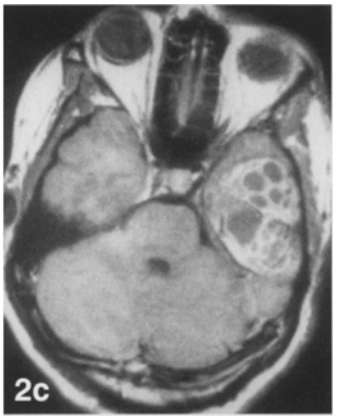


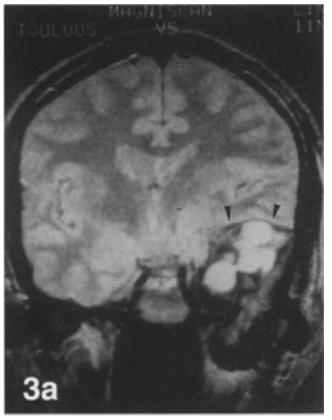
Fig. 1a, b. Contrast-enhanced T1 weighted axial MRI. A heterogeneous enhancing right mastoid tumour invades the posterior labyrinth, bordered medially by the dura mater of the posterior cranial fossa, without compromise of the lateral sinus

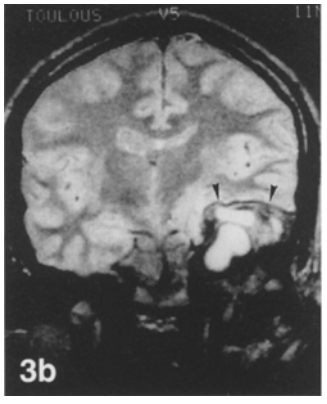
Correspondence to: A. Bonafé, Service de Neuroradiologie, Hôpital Purpan, Place du Docteur Baylac, F-31059 Toulouse Cedex, France











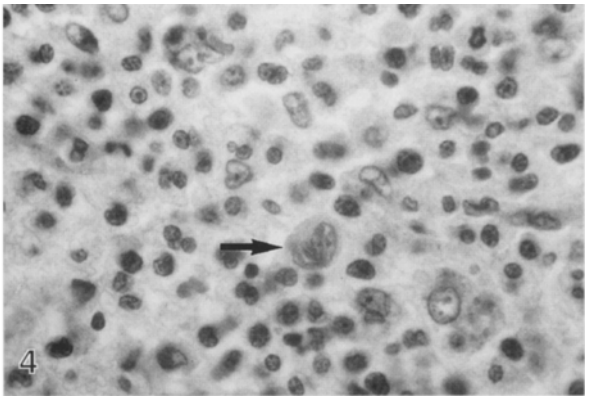


Fig. 2 a–c. T1-weighed axial MRI. **a** before, **b, c** after contrastmedium. A heterogeneous enhancing left petrous mass extends into the middle cranial fossa. Note cyst-like low signal areas with a thin contrast-enhancing rim

Fig. 3 a, b. T2-weighted coronal MRI. The cystic components of the tumour appear as rounded, high signal areas surrounded by incomplete low signal rims (possibly haemosiderin deposits). The extradural extension of the mass is bordered by an upward curvilinear displacement of the dura mater of the middle cranial fossa *(arrowheads)*

mogeneously. On T2-weighted images (Fig. 3), the tumour was isointense with the adjacent brain parenchyma. It displaced the temporal lobe anteriorly but remained extradural, as shown by a continuous low signal stripe representing the dura mater. The cystic areas gave high signal with T2 weighting, and were surrounded by a discontinuous low signal line representing calcification or haemosiderin deposits. Sagittal images showed that the tumour did not invade the intrapetrous segments of the internal carotid artery and jugular vein.

Total petrostomy, with monitoring of the seventh nerve was performed, with dissection of the intrapetrous segment of the carotid artery and the seventh nerve. Pathological examination (Fig. 4) showed in histiocytosis X. No other sites were involved (bone, lung, skin), and the patient received no futher treatment. Regular clinical and CT follow-up showed no evidence of recurrence.

Fig. 4. Histiocytosis X. Microscopic features of the granulomatous proliferation in which scattered cells of Langerhans type may be found *(arrow)*. (Haematoxylin and eosin)

Discussion

"Histiocytosis X" was proposed by Lichtenstein in 1953 as a unifying term under which three syndromes: eosinophilic granuloma [3], Hand-Schüller-Christian disease [4–6] and Letterer-Siwe disease [7, 8] were grouped. These three syndromes have distinctive symptoms and signs and prognosis, but are all characterised by abnormal proliferation of histiocytes. It therefore seems preferable to replace the three classical syndromes by a classification including the recent pathohysiological and histological data concerning localised and disseminated forms of histiocytosis X [2, 9, 10].

The localised form of histiocytosis X is commonly referred to as eosinophil granuloma, a term reserved for cases in which the disease is limited to bone or lung. Accounting for 60–80 % of cases of histiocytosis X [11], the localised form is the least aggressive, most favourable type [12]. Most of the patients with this form present between 5 and 10 years of age.

Alfred Hand first reported a yellow, fluctuant lesion, about the size of an American 5 cent coin, in the parietal bone [4, 13]. Several similar macroscopic descriptions of osseous histiocytosis X have subsequently been given. The lesions are brownish-yellow, with alternating soft tissue and cystic components. The cysts contain gelatinous or necrotic, haemorragic material. The abnormal histiocyte is a large cell, with abundant cytoplasm and granulations and inclusions in relation to its phagocytic properties. The nucleus contains a single nucleolus [14, 15] is irregular and lobulated, with longitudinal nuclear grooves. These cells form granulomas in conjunction with lymphocytes, eosinophils and polymorphs.

The electron microscopic characteristics of the abnormal histiocyte were described by Basset and Turiaf in 1965, and subsequently confirmed by others [11, 14, 16]. It consists of elongated intracytoplasmic structures, whose long axis is parallel to that of the all, with two symmetrical outer layers. The width of 420 Å is constant. Within the central axis, there are alternating dark and light bands approximately every 100 Å, the number, length and linear or curved configuration of which vary. These structures, called "zipper-like" or Burbeck granules, always present,

are necessary for the definitive diagnosis of histiocytosis X [9, 13, 17]. Apart from the macroscopic and ultrastructural features, the third key to the positive diagnosis is provided by immunohistochemical analysis [15], using specific markers (OKTG, S100 protein, HLADR), which are essential for differentiation of histiocytic from other polymorphic granulomas.

The aetiology remains unknown. A viral agent has been considered with the discovery of the Burbeck granules. However, this has never been proved, and various experiments consisting of inoculation to a living organism or to embryonic cells have not yielded positive results. Abnormal immune regulation is now considered most likely. Osband et al. [18] demonstrated an abnormal immune response in children with histiocytosis X and obtained clinical improvement after treatment with thymic extracts.

Ear, nose and throat (ENT) manifestations of histiocytosis X are rare. McCaffrey and McDonald [19], reviewing the literature between 1926 and 1978 suggested that ENT involvement occurs in about 15% of cases. Temporal bone involve even less frequent [11, 20]. Tos et al. [21] distinguish two clinical types of temporal bone involvement. In disseminated histiocytosis X, the temporal bone is involved in 61% of cases. Approximately 80% of the patients will experience ENT manifestations during the course of their disease. In localised osseous forms of the disease, isolated temporal bone involvement is very rare; only 19 cases have been reported [22]. Toohil et al. [23] observed only 16 cases between 1952 and 1972. Bouchez et al. [22] reported a woman aged 45 years a man aged 26.

The maximum incidence of isolated temporal bone involvement is between 3 and 10 years of age, mean $8^1/_2$ years [11, 24]. In the series of McCaffrey and McDonald [19] 59% of the patients were under 4 years of age. The sex ratio is the same as that for eosinophilic granuloma, i.e., a male-to-female ratio of 2:1 [25].

The clinical features can be purely otological or other manifestations of temporal bone involvement [22]. The otological manifestations include nonspecific external otitis (45%) and suppurative otitis media (59%), with a poor response to the usual treatments properly administered. Stenoses of the external auditory canal are due to extension to the posterior wall or to histiocytic granulation tissue within the canal.

Clinical manifestations of temporal bone involvement [1, 26] are present in 15–61% of cases, and are characterised by conductive deafness (via involvement of the ossicles or tympanic perforation) or perceptive deafness and vertigo. These are rare, as the osseous labyrinth is usually spared. Seventh nerve palsy, otorrhoea and local pain over the mastoid process are rare.

The typical lesion of osseous histiocytosis X of the skull is a lytic lesion without reactive sclerosis [15, 27, 28]. Petrous involvement is clearly demonstrated only by CT [20, 26, 29]. The lesions are lytic and without reactive sclerosis. The Fallopian canal and otic capsule are generally spared [31]. The soft tissue mass is isointense with adjacent brain but intense, homogeneous contrast enhancement [22, 26, 32, 33]. Contrast-enhanced CT permits assessment of subcutaneous and intracranial extension, skull base and vault invasion and intraparenchymal or pituitary stalk granulomas [32, 34, 35].

The MRI characteristics histiocytosis X localized to the petrous bone have not previously been reported. In our two cases, the MRI features correlated closely with the histopathological descriptions of soft tissue and cystic components, as described. T1-weighted MR images showed the lytic components within the enhancing tissue. MRI shows the byproducts of haemoglobin degradation found on the cyst walls (curvilinear low signal on T1 and T2-weighted images, representing haemosiderin deposits) which are to be differentiated from osseous remnants from incomplete osteolysis; comparison with the CT is useful. Furthermore, MRI reveals the absence of transdural extension and the patency of the vascular system.

Among the differential diagnoses of histiocytosis X of the petrous bone, only mastoid tumours will be considered. Rhabdomyosarcoma is often seen in children, representing the third most frequent childhood malignancy after leukaemia and neuroblastoma [19, 36, 37]. It accounts for 4–8% of malignant tumours in children under 15 years of age [38]. Up to 30–50% of rhabdomyosarcomas occur in the head, and 7% in the middle ear. This tumour of mesenchymal origin [39] derived from the pluripotent rhabdomyoblast, usually affects patients from birth to 20 years of age, mean 7.5 years; 30% are under 3 years of age. According to the intergroup study of rhadomyosarcomas (ISR) [40], the orbital is the most common site, with a good prognosis because of its early detection: 80–90% of survival after 5 years; parameningeal tumours (middle ear, maxilla and nasopharynx) have a poor prognosis because of their frequent extension to the skull base: 47% survival after 5 years; rhabdomyosarcomas in all other sites have a variable prognosis, the survival rate being 75% after 5 years.

In the case of parameningeal rhabdomyosarcoma with middle ear involvement, the symptoms and signs consist of recurrent otitis media with otorrhagea, seventh nerve palsy and hearing impairment; involvement of multiple cranial nerves is frequent. Cervical lymph node lung, pelvis, long bone and spinal metastases have been reported. CT is nonspecific, showing heterogeneous osteolytic lesions which enhance with contrast medium, more or less to the same level as adjacent muscle (41) but less so than histiocytosis X. MRI is also not very helpful in differential diagnosis. T1-weighted images show a hypointense lesion in the case of rhabdomyosarcoma of the middle ear [38], but assessment of bone destruction is more difficult; Latack et al. [38] considered CT superior to MRI for assessing extension of the disease.

In the adult, jugulotympanic chemodectomas must be considered in the differential diagnosis. Invasion and destruction of the petrous bone towards the apex and extension to the posterior cranial fossa via infralabyrinthine cells are observed [42]. T1-weighted MRI reveals a heterogeneous mass, isointense with cerebral parenchyma, and with scattered low signal areas representing blood vessels; the vascular nature of these images is confirmed on T2-weighted images [43].

When a primary tumour is known to be present, the diagnosis of metastases is easy but this diagnosis can be vir-

tually impossible when it is the first manifestation of the disease [44]. The preferred sites for metastases are the petrous apex and the internal auditory canal. The primary tumours are, in decreasing order of frequency, breast, lung, kidney, prostate, stomach, oropharynx, uterus and larynx [45, 46]; in the young child, adrenal neuroblastoma is the most common [45]. A few cases of multiple myeloma have been reported in children [47].

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