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Case Report

Eosinophilic Granuloma of the Temporal Bone in an Adult: Controversies in the Management

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A B S T R A C T

Eosinophilic granuloma is a rare, benign, lesion characterized by uncontrolled proliferation of Langerhan's cells. It is commonly found in a bone and has been described in almost every bone in the body. It usually affects children and is seldom found in adults. A case of 60-year-old adult male with an eosinophilic granuloma of the right temporal bone with infiltration of the temporal and infratemporal fossa and osteolysis of the squama of the temporal bone is presented. Diagnostic procedure and course of treatment are described. Controversies in the management and different approaches in therapy of such lesion are discussed because there is no agreed treatment protocol established.

Key words: *eosinophilic granuloma, Langerhan's cell histiocytosis, temporal bone, treatment modalities*

Introduction

Eosinophilic granuloma is a rare, benign, lesion characterized by uncontrolled proliferation of Langerhan's cells. It is commonly found in a bone and has been described in almost every bone in the body. It usually affects children and is seldom found in adults. Eosinophilic granuloma is subtype of Langerhan's cell histiocytosis (LCH), also termed histiocytosis X in the past. Eosinophilic granuloma was described and named by Lichtenstein and Jaffe in 1940[1–3]. In 1953, Lichtenstein proposed term histiocytosis X for a group of disorders characterized by proliferation of histiocytes[2–4] in different organs. In 1987 disorders under the term histiocytosis X were reclassified as Langerhan's cell histiocytosis (LCH)[5].

LCH encompasses three different subtypes: eosinophilic granuloma, Hand-Schüller-Christian disease and Letterer-Siwe disease. Eosinophilic granuloma is the most common form of LCH and accounts for 70% of all cases of LCH[6]. It is a solitary lesion, usually confined to skeleton with favourable prognosis. Solitary lesion can be also found in other organs like lungs or at some rare localisations like intraocular[7]. Hand-Schüller-Christian disease is characterized by multiple skeletal and visceral lesions, with symptoms of diabetes insipidus and less favourable prognosis. Letterer-Siwe disease has rapidly fatal course with involvement of multiple organs (skin, lymph nodes, spleen, lung, liver, brain and gastrointestinal tract) but without bone lesions[2,5].

Eosinophilic granuloma is found in children in 90% of cases[8]. Male to female ratio is 2:1[8]. It usually affects skull, vertebral spine and long bones, with skull being frequently involved in 30–80% of cases[6,9,10]. Ethiology is unknown. Proliferation of Langerhan's cells is monoclonal, so neoplastic process is possible. Autoimmune mechanism is also proposed; there is a report of case with co-existing eosinofilic granuloma with other autoimmune disease (psoriatic arthritis)[8]. Familial occurrence is rare in eosinphilic granuloma and Hand-Schüller-Christian disease but there is reported case of eosinphilic granuloma of the skull in identical twins[4]. Eosinophilic granuloma can be asymptomatic or can present with swelling, tenderness and localised pain.

In this article we present a case of an adult male with eosinophilic granuloma of the temporal bone. We also discuss different types of treatment modalities.

Case Report

A 60-year-old male was referred to Department of otorhinolaryngology head and surgery, complaining of localised swelling at the right temporal region for the last 1–2 months.

In past medical history he underwent tonsillectomy and appendectomy. He was taking therapy for type 2 diabetes and hypertension.

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The physical examination revealed mass at the right temporal region measuring approximately 5 cm (Figure 1). The mass was descending to the level of the zygomatic arch. On palpation, mass was soft and painless. No palpable lymph nodes were found in the region of right parotid gland and neck. Otomicroscopic examination of the right ear revealed normal tympanic membrane with normally shaped walls of the external auditory canal. Skull X-ray

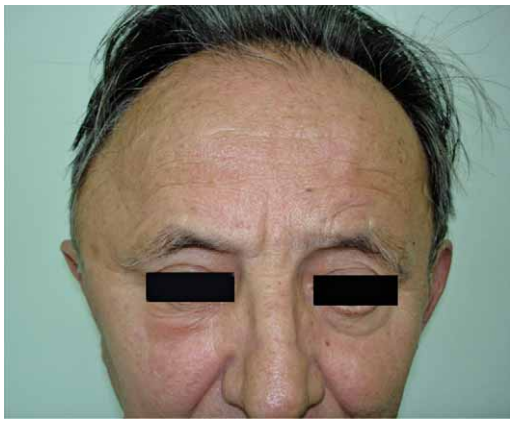

Fig. 1. Patient with a mass at the right temporal region.

revealed osteolytic lesion of the squama of the right temporal bone. Ultrasound of the right temporal region revealed mass measuring 40x50x13 mm with destruction of temporal bone measuring 12x6 mm. Fine needle aspiration was unspecific, diagnosis was mesenhimal tumour. Biopsy of the tumour was performed and histological examination revealed uniform histiocytes that were positive for CD-68, CD-1a and S-100 protein. These findings were consistent with LCH. Computed tomography (CT) showed solid mass in the area of right temporal fossa and superolateral part of infratemporal fossa with destruction of the squama of the temporal bone on two separate localizations. The lesion was in close proximity to dura so infiltration of dura could not be ruled out (Figure 2). Magnetic resonance imaging (MRI) confirmed presence of bone destruction and close proximity of tumour mass to dura but there were no obvious signs of dura infiltration (Figure 3). Scintigraphy of the whole

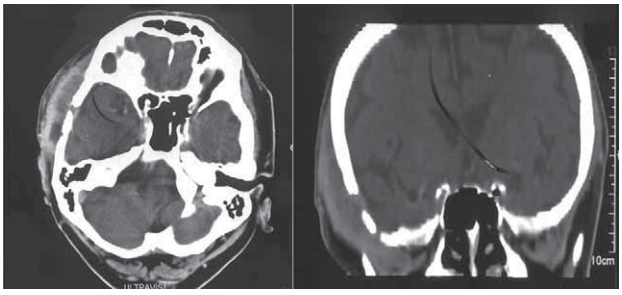

Fig. 2. CT (axial and coronal view) showing a mass at the region of right temporal fossa and infratemporal fossa with destruction of the temporal bone. The tumour is in close proximity to dura.

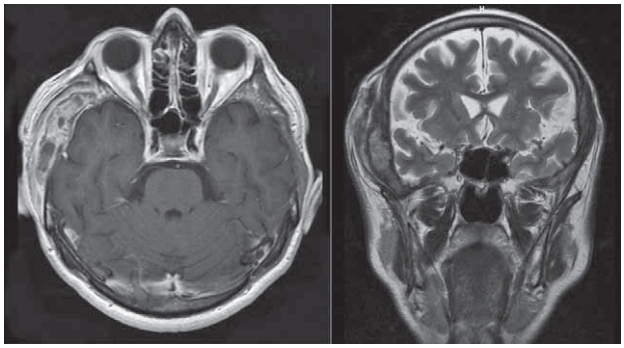

Fig. 3. MRI (axial-T1 signal and coronal-T2 signal) showing tumour with close relationship to dura but without obvious signs of dura infiltration.

body with Tc-99m didn't reveal any other bony lesions except of the right temporal bone. Chest X-ray was normal. Ultrasound of the abdomen didn't reveal any visceral lesions. Laboratory findings were within normal limits except slightly elevated erythrocyte sedimentation.

It was decided to surgically remove the tumour which included multidisciplinary team (maxillofacial surgeon, ENT and neurosurgeon). Hemicoronal approach was used to access the tumour. After removal of temporal muscle with tumour and periost of the temporal bone, destruction of temporal bone was found at four places, largest measuring about 2x1 cm and three smaller. Tumour was removed from the dura with preservation of dura in all four places and was also removed from superolateral portion of infratemporal fossa in piecemeal fashion (Figure 4). No removal of the bone was performed except localised curettage. Histological examination was consistent with findings on biopsy confirming LCH.

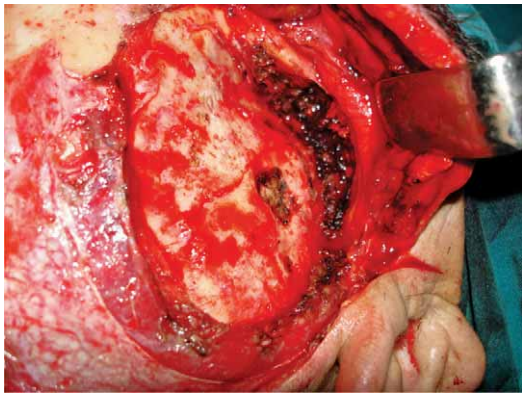

Fig. 4. Operation field after removal of the tumour. Destruction of the temporal bone is visible in the central part of the picture.

Patient recovered well after the surgery. Because of close proximity of tumour to the dura and not be able to achieve adequate margins without resecting the dura, patient was referred to adjuvant radiotherapy in consultation with oncologist.

Discussion

Eosinophilic granuloma is a rare tumour in adults and is more commonly found in children[3]. It is a solitary lesion which can affect almost every bone, but the skull is the most commonly site of involvement which was also the case in our patient. Diagnosis is made by histological examination and immunohistochemical staining. The pathologic Langerhan's cells are positive for CD-68, CD-1a and S-100 (which were also positive in our patient). On electron microscopy, Birbeck granules are seen within cytoplasm of the cells[5,8,10,11]. Differential diagnosis includes: giant cell reparative granuloma, aneurismal bone cyst, brown tumour, hematic cyst, osteomyelitis, fibrous dysplasia, rhabdomyosarcoma, Ewing's sarcoma, osteogenic sarcoma or metastatic lesion[3,5,10].

In management of solitary eosinophilic granuloma it is important to rule out multifocal or disseminated forms of LCH. It is accomplished by careful examination of other bones in the body and visceral organs by various imaging methods.

There are different treatment options described in publications. Some authors advocate no treatment because there are cases of spontaneous regression of LCH lesions[11]. Others advocate minimal surgical intervention like biopsy or curettage[3]. Some authors recommend surgical intervention followed by postoperative therapy that includes intralesional application of corticosteroids[5], postoperative low-dose radiotherapy[8] or combination of radiotherapy and chemotherapy[10]. In some cases only low-dose radiotherapy was applied[12]. Some authors advocate radical excision with wide margins[8]. Systemic therapy, like chemotherapy and systemic use of corticosteroids is used in cases of recurrence, progressive disease or appearance of multiple lesions[5,10]. There is opinion that radiotherapy is not necessary for treatment of solitary eosinophilic granuloma in childhood because patients treated without radiotherapy also showed improvement and because irradiation has its hazards, and it may induce iatrogenic tumours many years after therapy[3,5].

Local recurrence rate is 6% and new lesions appear in about 22% of patients[10], so long term follow-up is recommended.

In treatment of our patient we decided to surgically remove the tumour because it was accessible. Treatment included multidisciplinary approach (maxillofacial surgeon, ENT and neurosurgeon) because of destruction of bone and possible dura involvement. Postoperative radiotherapy was applied because of close proximity of tumour to the dura and not be able to achieve adequate margins without resecting the dura.

Our recommendation in treatment of solitary eosinophilic granuloma is surgical excision, without a radical approach, and with radiotherapy as adjuvant therapy if necessary. Radiotherapy should be avoided in children. Systemic therapy should be left for recurrence or appearance of multiple lesions.

Conclusion

Eosinophilic granuloma has favourable prognosis and its treatment should be individually. Treatment of eosinophilic granuloma depends mainly on localisation of the lesion and age of the patient. In the management of solitary eosinophilic granuloma, it is important to rule out multifocal or disseminated forms of LCH. Our recommendation is surgical excision, without a radical approach, and with radiotherapy as adjuvant therapy if necessary. Radiotherapy should be avoided in children. Systemic therapy should be considered in tumour recurrence or appearance of multiple lesions.

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EOZINOFILNI GRANULOM TEMPORALNE KOSTI U ODRASLOJ DOBI: DILEME U LIJEČENJU

S A Ž E T A K

Eozinofilni granulom je rijetka, dobroćudna, lezija karakterizirana nekontroliranim rastom Langerhansovih stanica. Često je smještena u kostima te je opisana u skoro svim kostima u tijelu. Uobičajena je pojava bolesti u dječjoj dobi dok se rijetko pojavljuje u odrasloj dobi. U ovom radu prikazujemo slučaj odraslog muškarca, u dobi od 60 godina, sa eozinofilnim granulomom desne temporalne kosti, zahvaćanjem temporalne i infratemporalne jame i probojem ljuske temporalne kosti. Opisujemo dijagnostičke metode i način liječenja. Obrazlažemo dileme i različite pristupe u liječenju jer ne postoji usuglašeno stajalište o liječenju ove bolesti.