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Case report

Langerhans' cell histiocytosis of the mandible in bone scintigraphy and CT

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Abstract

The authors present a case of a 4-year old boy with a quickly growing tumor of the jaw. The CT examination revealed a destructive tumor in the body of the mandible involving soft tissues. A diagnosis of eosinophilic granuloma of the mandible was confirmed by a biopsy of the tumor. Skeletal scintigraphy showed areas of increased and decreased radiotracer uptake. The fusion of CT and scintigraphy images showed that the "cold" focus corresponds with the osteolytic area and the "hot" focuses are larger than the areas of osseous reconstruction shown in CT. Conclusion: In cases of histiocytosis skeletal scintigraphy and CT are complementary methods that enable one to make an assessment of the extent of the disease.

Key words: Langerhans' cell histiocytosis, scintigraphy, CT

Introduction

Eosinophilic granuloma is one of the forms of Langerhans' cell histiocytosis that occurs in children predominantly between 2 and 10 years old. Most often a single osteolytic focus in flat bones, vertebrae and long bones is found. Histopathological examination shows the typical mixture of Langerhans' cells and eosinophilic granulocytes.

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Case description

A 4-year old boy was admitted to the hospital because of a quickly growing, firm, immobile and painful during palpation tumor in the mental area. The patient had been suffering from increasing anxiety, sleep disorders, pain and swelling of the mental area for about 1 month.

During physical examination a bulged outline of the body of the mandible, with a swelling of soft tissues was found. These complaints were accompanied by heat and tenderness of the mental area with enlarged, painless lymph nodes of the submandibular and sterno-cleidomastoid muscle area. In laboratory findings an increased activity of alkaline phosphatase: 423 U/l (norm: 0–270), hyperphosphataemia: 1.89 mmol/l (norm: 0.87–1.45) and hypocalcaemia: 1.96 mmol/l (norm: 2.15–2.55) were found.

No abnormalities were shown during a chest X-ray and abdominal ultrasonography. No bone lesions were found during an X-ray examination of the skull, pelvis and long bones of the extremities.

During ultrasonography of the mental area an irregular, hypoechogenic focus of 15 mm × 8 mm size was revealed which was thought to be a tumor. Some small hypoechogenic focuses about 6 mm in size, suggesting lymph nodes, were found near the tumor.

A CT examination was performed before and after the administration of a contrast medium. In the body of the mandible a pathological mass of 41 *H.U.* and 50 *H.U.* density was visualized. The tumor caused mandibular osteolysis within the range of 28 mm × × 35 mm × 34 mm (about 16 cm^3). The pathological mass infiltrated the soft tissues of the bottom of the oral cavity (Figure 1). Tooth buds, that were localized within the pathological process, were not affected. Along the sterno-cleidomastoid muscles and in the area of the mandible angles, several single lymph nodes up to 10 mm and 14 mm were found.

During a skeletal scintigraphy, two areas of pathological radiotracer accumulation were shown in the area of the mental protuberance. No other abnormalities of the skeletal system were found (Figure 2).

The diagnosis of Langerhans' cell histiocytosis was confirmed on the basis of a histopathological examination of the material taken during an open biopsy of the tumor. A trepanobiopsy

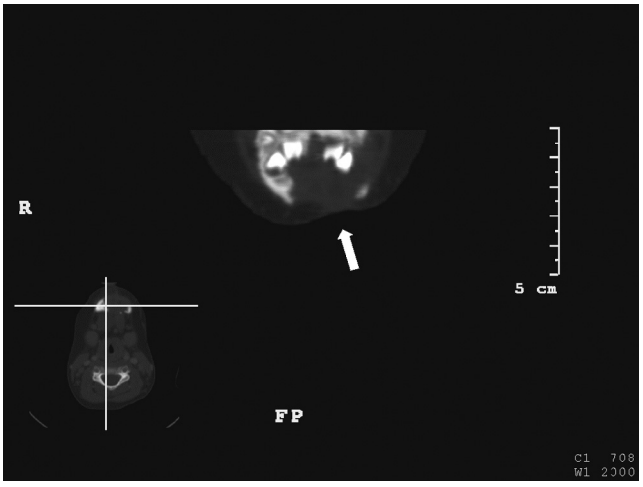

Figure 1. Computer tomography. Frontal reconstruction (MPR). Osteolysis of the mandible and a tumor within soft tissues (arrow).

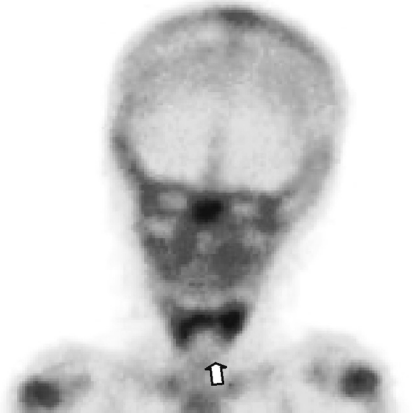

Figure 2. Skeletal scintigraphy. Anterior projection. Increased radiotracer accumulation in the body of the mandible with "cold" focus in the mental area (arrow).

revealed: bone marrow rich in cells, no histiocytic infiltration focuses, all the haemopoietic lines represented.

Discussion

Langerhans' cell histiocytosis (LCH), also called histiocytosis X, is a rare disease that occurs predominantly in children between the ages of 2 and 10. The Frequency of LCH is estimated at 2–5 cases per 1 mln per year [1, 2]. There are 3 main types of LCH: Letterer-Siwe disease, Hand-Schiller-Christian disease and eosinophilic granuloma [2, 3]. The common pathological feature of these 3 types of the disease is an accumulation of dendritic cells that originated from the bone marrow stem cell, from the monocyte-macrophage's line [4]. The proliferation of histiocytes is often accompanied by an infiltration of multinucleated giant cells and eosinophils [5]. Distinguishing between the above-mentioned types of LCH may be difficult and actually LCH is categorized into unifocal (single organ involvement) and multifocal disease [6, 7]. Besides the skeletal system LCH may affect the lungs, liver, spleen, gastrointestinal tract, kidneys, central nervous system, posterior pituitary gland, thyroid gland, parathyroid glands, thymus, skin, mucosal membrane and lymph nodes [2, 6, 8, 9–11].

In the presented case of the 4-year old boy, lesions were located in the body of the mandible and surrounding soft-tissue especially in the inner soft tissue of the oral cavity. Di Nardo and Wetmore in their studies estimated that the incidence of head and neck complaints exceeded 80% of cases [12]. Irving et al. report that the skull is the most frequently involved area within head and neck locations. The pathological process affects in decreasing order of frequency cervical nodes, temporal bone, maxilla and mandible [13]. Mandible involvement is found in 5.0–5.5% of cases [11].

In the described case, the diagnosis of LCH was established on the basis of histopathological examination of the material taken during an open biopsy of the tumor. The diagnosis of disseminated type of Langerhans cell histiocytosis was established with regard to the mandible and surrounding soft tissue involvement. The boy was qualified for the lower risk group — patients over the age of 2 years, without the involvement of the haematopoietic system, liver, lungs and spleen.

The prognosis of LCH depends particularly on the number of organs involved. Thus, accurate identification of pathological lesions is very important. Therefore, radiography and CT of suspected areas are used routinely [3]. Some lesions with no clinical manifestation may not be detected by these techniques. In patients with bone involvement the skeletal scintigraphy has been shown to be complementary to conventional radiography [11, 14]. In the described case the extent of the pathological process was established on the basis of CT and scintigraphy examinations. During CT examination with a three-dimentional reconstruction and a 4D presentation (Figure 3), an irregular osteolytic focus with cortical layer disruption in the body of the mandible was found. The bone lesion was accompanied by a pathological mass within the soft tissues without calcification and ossification. No osteolytic lesions were visualized within the deciduous and permanent tooth buds. During the skeletal static scintigraphy after the administration of 400 MBq ^{99m}Tc-MDP areas of increased and decreased radiotracer accumulation in the body of the mandible were shown. No other abnormalities of the skeletal system were found. The fusion of CT and scintigraphic images showed that the "cold" focus corresponds with osteolytic area and "hot" focuses are larger than the areas of osseous reconstruction visualized in CT (Figure 4).

Some authors point out that CT is the preferred method for assessing disease extension in the skeletal system, monitoring of disease activity and assessing the response to treatment [11, 15]. A bone scintigraphy can have a role in the diagnosis and staging of LCH [3, 11, 15]. Howarth et al. reported that the sensitivity and specificity of radiographic survey for the detection of LCH were 100% and 61%, respectively, compared to 91% and 55% for bone scintigraphy. For the detection of LCH the accuracy of a skeletal survey was 90% compared to 82% for bone scintigraphy [11]. Howarth et al. reported also that for the detection of multifocal

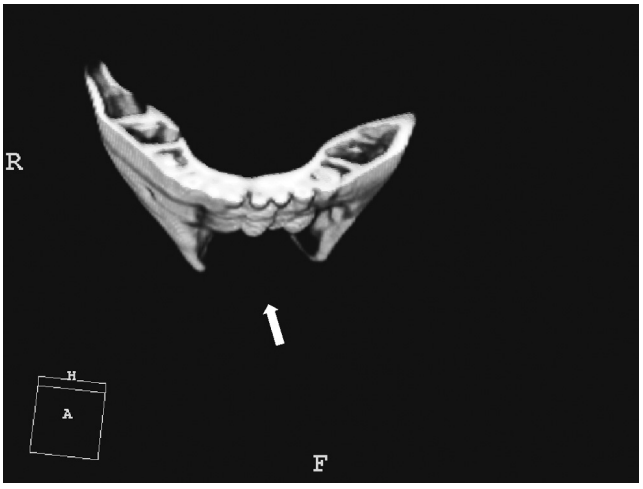

Figure 3. Computer tomography. 4D reconstruction. Osteolysis of the mandible (arrow).

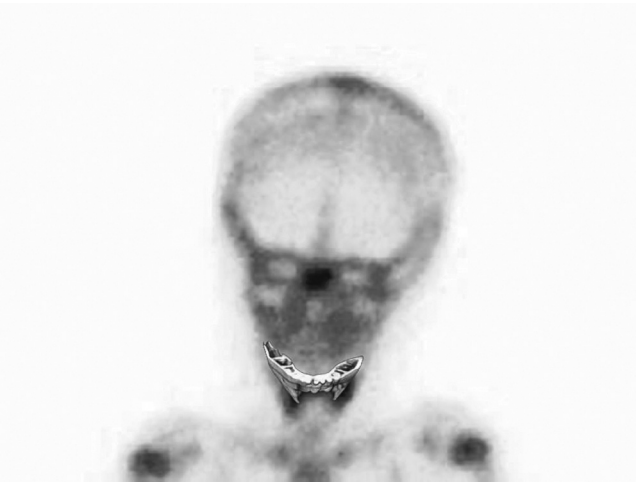

Figure 4. The fusion of CT and scintigraphic images (4 D reconstruction) — anterior projection.

osseous LCH lesions, radiography is the preferred imaging method. Bone scintigraphy can be a reliable diagnostic tool for the detection of solitary osseous lesions. For solitary lesions, radiograph sensitivity was 95% compared to 88% for bone scintigraphy [11]. A regional analysis of the bone scintigraphy showed poor sensitivity in the detection of LCH lesions localized in the pelvis, sacrum, ribs, sternum, clavicles and scapula. The skull, facial bones and mandible sites showed a higher sensitivity, but a lower specificity [11]. Dogan et al. report that bone scintigraphy is more sensitive than X-ray skeletal examination. In his study of the 191 lesions detected in 42 patients, 36 (19%) were not detected during scintigraphy and 55 (29%) were missed during X-ray examinations. Most of the undetected lesions during scintigraphy were located in the skull (26 of 36). Thirteen skull lesions were not detected during X-ray examination, but seen during scintigraphy. Most of undetected lesions during X-ray examination were located in the ribs (20 of 30 rib lesions), whereas during scintigraphy 29 of them were identified [14].

There are some trials using radiopharmaceutical agents with an affinity to somatostatin receptors for the identification of histiocytic lesions [3, 16, 17]. Weinmann et al. reported that 111-In-pentetreotide (Octreoscan) scintigraphy may be a complementary method to CT and skeletal radiography, enabling the visualization of numerous involved sites in a single examination [3].

Conclusions

1. In the case of Langerhans' cell histiocytosis (LCH) skeletal scintigraphy and CT are complementary examinations enabling an assessment of the extension of the disease.
2. The fusion of scintigraphy and CT images enables one to make an assessment of a single pathological focus more precisely.

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