Case Report / Olgu Sunusu

Diagnostic Imaging of Lymphangiomatosis- A Pictorial Overview

Lenfanjiomatozisin Tanısal Görüntülemesi – Resimlerle Genel Bakış

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Başvuru Tarihi/Received :

10-05-2012

Kabul Tarihi/Accepted:

21-05-2012

ÖZET

Lenfanjiomatozis; kemikler, parankimal organlar ve yumuşak dokuyu kapsayan lenf kanallarının diffüz proliferasyonu ile karakterize nadir görülen konjenital bir bozukluktur. Lenfanjiomatozis sıklıkla geç çocuklukta görülür. Birden çok kemiklerde litik lezyonlar oluşturur. Hastalığın prognozu iyi huylu durumdan ölümcül hale kadar çeşitli şekillerde rapor edilmiştir. Biz özellikle mandibula tutulumu olan lenfanjiomatozisli 14 yaşındaki bir kız klinik ve görüntüleme bulguları sunuldu.

Anahtar Kelimeler: lenfanjiomatozis, mandibula, lenfanjiyom

ABSTRACT

Lymphangiomatosis is a rare congenital disorder, characterized by a diffuse proliferation of lymphatic channels involving bones, parenchymal organs, and soft tissue. Lymphangiomatosis frequently presents in late childhood. It produces lytic lesions in multiple bones. Prognosis of the disease has been variously reported from benign to fatal. We present the clinical and imaging features of a 14-year old girl with lymphangiomatosis and particularly involvement of mandible.

Key Words: Lymphangiomatosis, mandible, lymphangioma

Introduction

Lymphangiomatosis is a rare disorder characterized by diffuse proliferation of lymphatic channels involving bones, parenchymal organs, soft tissue, which occurs mainly in children and young adults.[1] Generalized lymphangiomatosis can involve any organ except the central nervous system, and the prognosis depends on the involving organ and the extent of the disease.[2] The exact incidence of pure lymphangiomas involving the skeleton is not well documented. [3] This is due to the wide range of clinical features and presentations of this rare entity.[4] Due to the rarity of the disease features of lymaphangiomatosis is not well described. Because radiologic examination is useful for evaluating the morphologic feature as well as the extent of disease, it is important to know the radiologic findings of lymphangiomatosis. In this report, we describe the radiologic findings of lymphangiomatosis in the cervical soft tissues, skull as well as the mandibular condyle, which is relatively rare site.

Case report

A 14-year-old girl reported to us with complaint of swelling on the right side of face and neck, of two year duration and gradual progression. Swelling was diffuse in nature, soft and nontender on palpation. Orthopantomograph revealed ill defined radiolucency and expansion of the right condylar neck and head region (Figure 1).

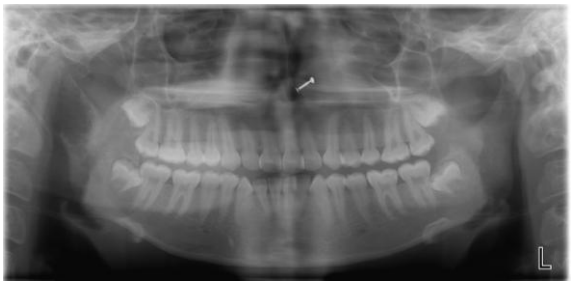

Figure 1 : Orthopantomograph showing ill defined radiolucency and expansion of the right condylar neck and head region

Postero-anterior skull view revealed multiple osteolytic areas in the skull vault (Figure 2). MRI performed to delineate the extent of the cervical lesion revealed large irregular cystic lesions in the neck involving multiple spaces

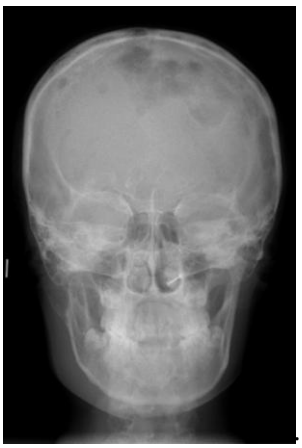

Figure 2 : PA skull view showing multiple osteolytic areas in the skull vault and expansion of right mandibular condyle

(Figure 3, 4). Septations were seen in the lesion. Several disseminated lesions were seen in mandibular condyle, skull, vertebrae, clavicle, ribs and sternum (Figure 4,5,6). Histopathological specimen obtained from parotid region revealed features of lymphangiomatosis. Due to wide spread extension of lesion and lack of expertise at the local level, patient was referred to higher centre for evaluation and treatment. Patient was under continuous follow up for the period of one year. Surgical excision of the cervical lesion was carried out at higher centre. The lesions present in the mandible were followed up through panoramic radiographs. There was no increase in the right condylar lesion. Patient was referred back to the higher center for the further evaluation.

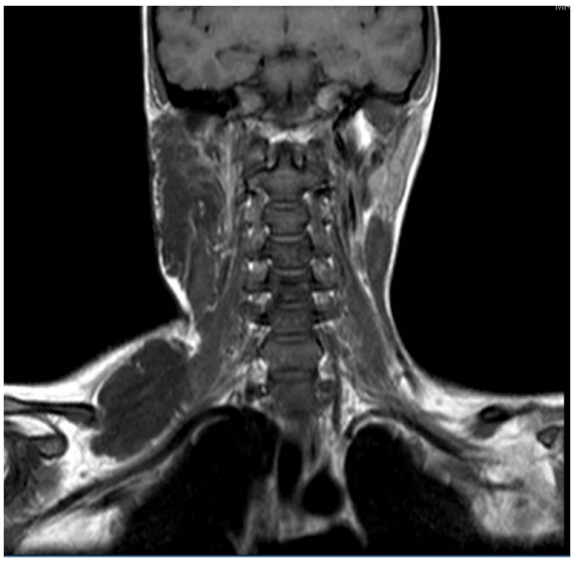

Figure 3: Coronal T1WI shows that the parotid gland is replaced by the abnormal low signal intensity lesion. The abnormal signal extends into the superficial neck spaces and the root of neck.

Discussion

Lymphangiomatosis is a benign malformation of the lymphatics characterised by abnormal proliferation of the lymphatic channels.[4] It was first described by Rodenber in 1828.[5] It is a congenital disorder with slow growth and multisystem involvement affecting the visceral and/or skeletal system, which results in a wide spectrum of clinical features. Histologically, there are three categories: capillary or simple, cavernous, and cystic.[4] Lymphangiomatosis has been described in patients ranging from birth up to 80 year. It most frequently presents in late childhood.[6] There is no clear sex predilection. We noticed in a 14 year old girl. The lesions of lymphangiomatosis can occur in any tissue in which lymphatics are normally found, but there appears to be a predilection for thoracic and neck involvement as observed in our case.[5] Most lymphangiomas present with a mass or diffuse swelling, which initially grows slowly with the child and after some time might regress. The symptoms depend on the sites of involvement.[7] It produces lytic lesions in multiple bones.[8] Lytic lesions in the cranial vault, mandible, vertebrae, clavicle, ribs, sternum were observed in our case. Cases of lymphangiomatosis affecting mandible have rarely been reported.[1] The usual symptoms that patient present depends upon the organs involved, such as chest tightness, breathlessness when thorax is involved and cosmetic abnormality when maxillofacial region is involved as seen in our case.[9]

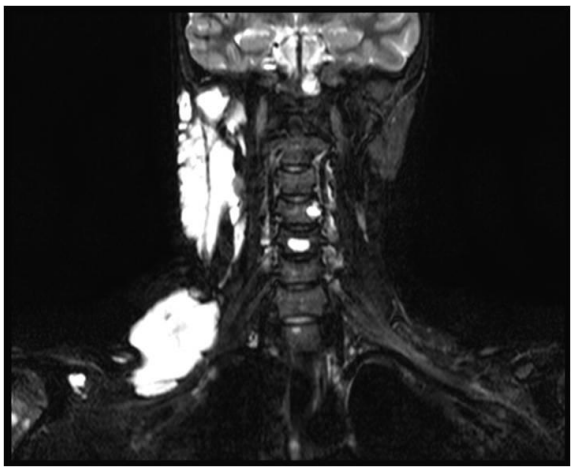

Figure 4: Coronal STIR images demonstrates the lesion as areas of high signal intensity areas. The involvement of the vertebral bodies is also shown.

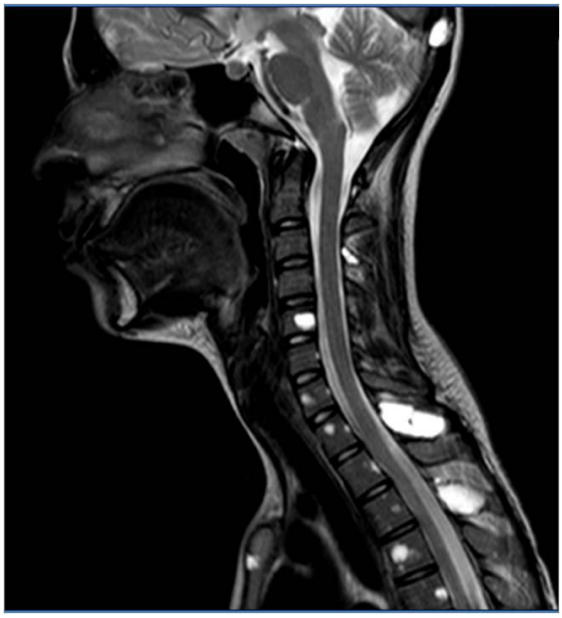

Figure 5: Sagittal T2WI shows involvement of vertebral bodies, sternum, skull vault as well interspinous ligaments.

The characteristic radiographic appearances of the bony lesions have been described as radiolucent areas with fairly narrow zones of transition. A faint, thin rim of sclerosis may be present. The bony lesions often start in the marrow but may be confined to the cortex. There is neither a periosteal reaction nor adjacent soft tissue swelling.[4] We noticed a ill defined radiolucency in the right condyle and multiple osteolytic areas in the skull vault in the plain radiographs. On CT these lesions are hypodense, nonenhancing, confluent low attenuation masses. On MRI these lesions show variable signal intensity. They are well-defined hypo to isointense on T1weighted images. On T2 weighted, STIR (short tau inversion recovery) and gradient echo pulse sequences the lesions appear hyperintense.[10] Similar features were noted in our case.

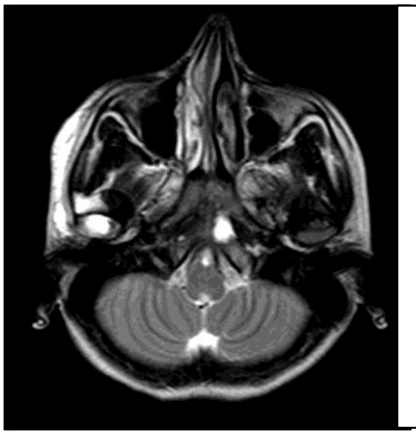

Figure 6: Axial T2weighted image showing high signal intensity in the right mandibular condyle and involvement of the clivus .

This disease may show a benign cause in some patients and cause serious morbidity in others.[8] Sometimes, because of their size and localization, the therapeutic approach to these lesions may become very challenging. Due to the rarity of the disease clear treatment strategies have not yet emerged. Several methods are used in literature, including surgical excision, sclerotherapy with bleomycin or OK-432, *α*-interferon therapy, treatment with Nd-YAG laser or percutaneous cementoplasty, and treatment with bisphosphonates, radiotherapy, chlorambucil/etoposide, and methylprednisolone. Follow-up of the patients with lymphangiomatosis was also proposed as an alternative management strategy in selected patients.[7] Surgical excision of the cervical lesion was carried out for our patient and she was followed up regularly. The aim of this report was to present occurrence of lymphangiomatosis at a relatively rare site of mandibular condyle.

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