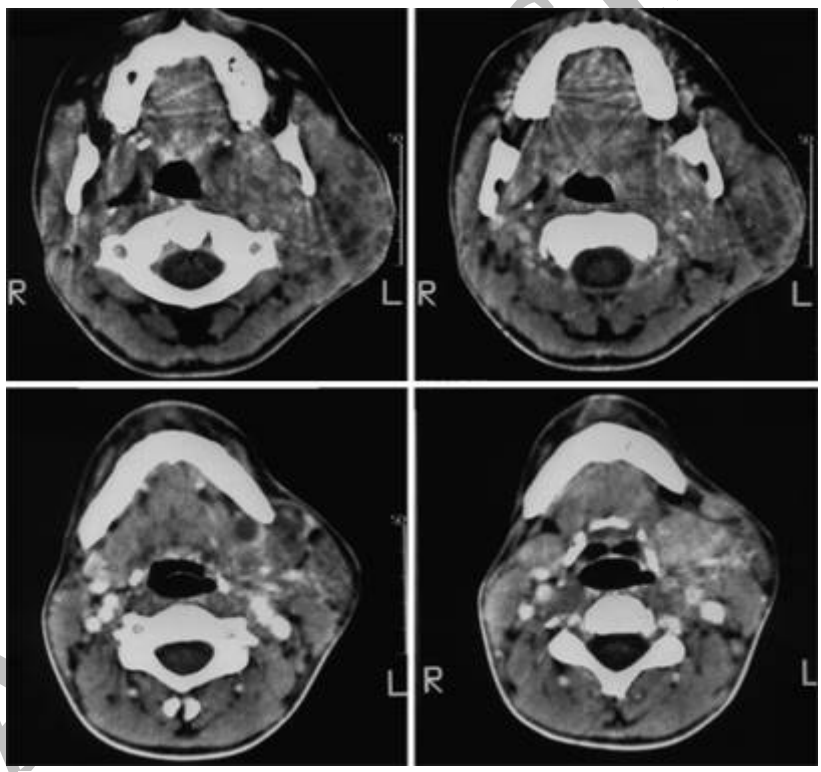
What is your diagnosis?

A 15-year-old female was referred with gradual painless swelling of her left parotid gland since 6 months ago.



Figs. 1-4. Show enlargement of the left parotid gland accompanied with multiloculated poorly differentiated fluid containig areas and intervening septa with extension to the parapharyngeal space and submandibular gland.

What is your diagnosis?

Diagnosis: Lymphangioma

L. Aghaghazvini MD[1]
H. Mazaher MD[1]
H. Sharifian MD[1]
Sh. Aghaghazvini[1]
1. Department of Radiology, Amir-Aalam Hospital, Tehran University of Medical Sciences, Tehran, Iran.

Corresponding Author: Leila Aghaghazvini
Email: la_ghazvini@yahoo.com
Tel: +9821-66704998
Fax: +9821-66704998

Lymphangiomas are rare congenital malformations, commonly seen in the head and neck region in 90% of cases.[1] They are uncommon, hamartomatous, congenital malformations of the lymphatic system that involve the skin and subcutaneous tissue and account for 4% of all vascular tumors and approximately 25% of all benign vascular tumors in children. The most common sites are the head and neck, followed by the proximal extremities, the buttocks, and the trunk. However, they sometimes can be found in the intestines, pancreas, and mesentery. They classically occur in association with congenital malformations of the lymphatic, presenting in childhood as cystic masses (cystic hygroma). They may also occur in adults in different locations including the oral cavity, tongue, larynx or parotid.[1-4] The classification most frequently used divides these lesions into two major groups based on the depth and the size of these abnormal lymph vessels. The superficial type is called lymphangioma circumscriptum. The deeper-seated group includes cavernous lymphangioma and cystic hygroma. Cavernous lymphangioma is also uncommon and usually arises during infancy. The most common sites are the head and neck areas and less frequently, the extremities. These lesions are seated deep in the dermis, forming a painless swelling or thickening of the skin, mucous membrane, and subcutaneous tissue.[1] They are usually infiltrative lesions that do not respect the fascial boundaries (trans-spatial disease) and may extend into multiple spaces.[5] They have no risk of malignant transformation but with a strong tendency for local recurrence unless they are completely excised. Recurrent episodes of cellulitis and bleeding are not uncommon.[1,3]

On imaging, they typically appear as multiloculated, poorly circumscribed lesions with intervening septation in CT and MRI. Enhancement of septations can occur after contrast administration, however most of the lesions do not enhance. On MRI these masses follow water signal with hypo intense T1-weighted and hyper intense T2-weighted signal to muscle. As a result of hemorrhage they may present as fluid-fluid level.[5]

The preferred treatment for lymphangioma is complete surgical excision. Adequate excision of lymphangioma may be difficult and this is the main reason for the high recurrence rate.[4]

This case is a 15-year-old female referred with a history of gradual painless swelling of the left parotid gland and constant left sided pharyngeal discomfort since 6 months ago. On physical examination, a rubbery enlarged parotid gland without any discrete mass or change in skin color was seen. Examination of the oropharynx revealed marked unilateral protrusion of the left tonsil, which was firm on palpation. Computed tomography revealed enlargement of the superficial part of the left parotid gland with multiloculated poorly differentiated fluid containing areas with intervening septation which extended medially to the deep lobe of the parotid gland accompanied with effacement of left parapharyngeal space and bulging of left tonsilar fossa. Extension of the lesion to the left submandibular gland and space was also seen. Adjacent subcutaneous fat appeared normal. No adenopathy was seen. According to the above mentioned findings, the location of the lesion, the manifestations and finally the patients age, lymphangioma, lymphoma and myoepithelial sialadenitis are mentioned. Hemangioma should also be considered with lower possibility. Considering the typical multi-

loculated, poorly circumscribed lesion with fluid attenuation and intervening septation, multiple space involvement with respect to subcutaneous fat planes and unilateral presentation of the lesion with no evidence of adenopathy, lymhangioma was our first diagnosis. The lesion was easily excised surgically without any adhesion and invasion to adjacent structures.

Pathology confirmed our diagnosis, which was lymphangioma of the parotid gland with extension to adjacent spaces.

References

1. Davis C Jr. Synchronous cystic hygromas. IMJ Ill Med J 198 Nov 8;174(5):292-4.
2. Friedberg J. Pharyngeal cleft sinuses, cysts and other benign neck lesions. Pediatr Clin North Am 1989 Dec;36(6):1451-69.
3. Williams HB. Hemangiomas and lymphangiomas. Adv Surg 1981;15:317-49.
4. Stal S, Hamilton S, Spira M. Haemangiomas, lymphangiomas and vascular malformations of the head and neck. Otolaryngol Clin North Am 1986:19:769-96.
5. Haaga Jo. CT and MRI imaging of whole body; vol 1. 4th edition. Ohio: Mosby; 2003. p. 648-649, 714.