Agenesis of isthmus of thyroid gland, its embryological basis and clinical significance – A case report

GP Kumar,[1] N Satyanarayana,[2] N Vishwakarma,[3] R Guha,[2] AK Dutta[2] and P Sunitha[4]

[1]Department of Anatomy, Sri Muthukumaran medical college Hospital and Research Institute, Chikkarayapuram Chennai, India, [2]Department of Anatomy, College of Medical Sciences, Bharatpur, Chitwan, Nepal, [3]Geetanjali Medical College and Hospital, Udaipur, India, 4Department of Physiology, College of Medical Sciences, Bharatpur, Chitwan, Nepal

Corresponding author: Dr. G. Praveen Kumar. MD (Anatomy), Assistant Professor, Department of Anatomy, Sri Muthukumaran Medical College Hospital and Research Institute Chikkarayapuram Chennai- 600069, India; e-mail: praveengattu@yahoo.com

ABSTRACT

A wide range of morphological varieties and developmental anomalies of the thyroid gland have been reported in literature such as hypoplasia, ectopy, hemi agenesis, and agenesis. Out of these the incidence of agenesis of the isthmus of thyroid gland is rare and very few cases have been reported. In the present case report a male cadaver was found with agenesis of isthmus of thyroid gland with pyramidal lobe and levator glandulae thyroidae arising from right lobe.

Key words: Agenesis of isthmus, pyramidal lobe, levator glandulae thyroidae.

The thyroid gland is brownish-red and highly vascular, is placed anteriorly in the lower neck , level with the fifth cervical to the first thoracic vertebrae, ensheathed by the pretracheal layer of the deep cervical fascia, it has right and left lobes connected by a narrow, median isthmus. Lobes are approximately conical, their ascending apices diverge laterally to the level of the oblique lines on the laminae of the thyroid cartilages, their bases are level with the fourth or fifth tracheal cartilages, each lobe is about 5cm long, its greatest transverse and anterio posterior extents being about 3cm and 2cm respectively, its posteromedial aspect is attached to the side of the cricoid cartilage by a lateral thyroid ligament.

Isthmus connects the lobes lower parts, it is about 1.25cm transversely and vertically and is usually anterior to the second and third tracheal cartilages though often higher or sometimes lower, its site and size vary greatly. The superior thyroid arteries anastomose along its upper border, at the lower border the inferior thyroid veins leave the gland.

A conical pyramidal lobe often ascends towards the hyoid bone from the isthmus or the adjacent part of the either lobe, it is occasionally detached or in two or more parts a fibrous or fibromuscular band.

The levator of the thyroid gland- muscular levator glandulae throidae, sometime descends from the hyoid body to the isthmus or pyramidal lobe.[1]

CASE REPORT

During routine midline dissection of the neck in the Department of Anatomy, Sri Muthukumaran medical college hospital and research institute Chennai a male cadaver showed agenesis of isthmus, pyramidal lobe and levator glandulae throidae was seen arising from the right lobe of thyroid gland. The thyroid gland had two separate lobes, with complete agenesis of isthmus.

Each lobe is pyramidal in shape and present apex and base, the length of the lobes was 6 cm at the right and 6.3 cm at the left lobe; widths were 3.3 and 3.9 cm, respectively.

Apex is directed towards the oblique line of thyroid cartilage and related to superior thyroid artery and external laryngeal nerve, the base is extended up to 5th, 6th tracheal rings, the base is related to inferior thyroid artery and recurrent laryngeal nerve. The location of thyroid gland was normal. The two lobes were separate without any tissue intervening between them (Figure 1). Normally isthmus connects the lobes lower part, it is about 1.25cm transversely and vertically and is usually anterior to the second and third tracheal cartilages though often higher, in the present case it is absent.

The pyramidal lobe of thyroid gland is remnant of caudal end of the thyroglossal duct and in the present cadaver it arose from the right lobe of the thyroid gland.

Each lateral lobe of thyroid gland were supplied by branches of superior and inferior thyroid arteries. Their was no abnormality found in the blood supply of the gland. No accessory thyroid arteries were present.

DISCUSSION

Agenesis of thyroid isthmus is reported by earlier anatomist and embryologist.It can be explained in the light of embryological development .Reports in the

Fig. 1. Showing agenesis of isthmus of thyroid gland.

RCCA=Right common carotid artery,LCCA=Left common carotid artery,LGT=Levator grandule thyroideae,LLT=Left lobe of thyroid gland,RL

T=Right lobe of thyroid gland, PL=Pyramidal lobe,TC=Trachea

literature suggest that chromosome 22 play role in the thyroid development.[2] Normally the two lobes of thyroid gland are joined together by an isthmus in the upper part of trachea.

The thyroid gland appears as an epithelial proliferation in the floor of the pharynx between tuberculam impar and the copula at a point later indicated by foramen cecum. Subsequently, thyroid gland decends in front of the pharyngeal gut as bilobed diverticulum. During this migration, the thyroid remains connected to the tongue by a narrow canal, the thyroglossal duct, this duct normally disappears.[3]

Rarely, a high separation of thyroglossal duct can engender two independent thyroid lobes and pyramidal lobes with the absence of isthmus.[4] Further development, the thyroid gland decends in front of the hyoid bone and the laryngeal cartilages. It reaches its final position in front of the trachea in the seventh week. By then it has acquired a small median isthmus and two lateral lobes.

In our case, the agenesis of isthmus of thyroid gland was noted may be due to a high separation of thyroglossal duct can engender two independent thyroid lobes and pyramidal lobe with the absence of isthmus. Further observation reveled that it was not associated with other anomalies of gland, and it may be a congenital anomaly.

The isthmus may be missing in amphibians, birds and among mammals - Monotremes, certain Marsupials, Cetaceans, Carnivores and Rodents. In rhesus monkey (Macacus rhesus), the thyroid glands are normal in position but there is no isthmus.[5]

Small detached masses of thyroid tissue may occur above the lobes or isthmus as accessory thyroid glands, vestiges of thyroglossal duct may persist between the isthmus and the foramen caecum of the tongue, sometimes as accessory nodules or cysts of thyroid tissue near the midline or even in the tongue.[1]

In litrature absence of Isthmus of the thyroid gland was also reported in 1952 by Allan. He states that isthmus was absent in 2.0 to 4.0% of cases. He also observed that a band of connective tissue named levator glandulae thyroidae extended from the apex of right or left lobe or isthmus of the thyroid gland to the hyoid bone. [6]

The variations in the size, shape and variation in the level of the thyroid gland was reported by Anson in 1996. He states that isthmus was absent in 6.0 to 8.0% of cases.[7]

The presence of pyramidal process was also found in 43.0% cases was reported by Marshall in 1975. The pyramidal process was sometimes found connected to the isthmus and to one of the lobes.[8]

Eisler 1922 made an extensive study on the leavator glandulae thyroidae and its innervations. He states that the levator of the thyroid gland may be innervated either by ansa cervicalis or through vagus. He has also regarded the levators of the thyroid gland are divisible into three groups viz., the anterior, lateral and posterior levators derived from cricothryoid, infrahyoid and inferior constrictor muscle of pharynx.[9]

The morphological difference in the evolutionary origin does not result in any changes in thyroid function. Usually agenesis of isthmus is difficult to determine unless the patients refer for other thyroid diseases.

Agenesis of isthmus can be diagnosed via scintigraphy, ultrasonography, CT and MRI. When absence of isthmus is suspected, the individual may be directed for a differential pathological diagnosis such as autonomous thyroid nodule; thyroiditis; primary carcinoma; neoplastic metastases; and infiltrative diseases such as amyloidosis.[10]

Under standing of thyroid anatomy and associated anatomical variations are very important so that these variations are not over looked in the differential diagnosis.

REFERENCES

1. Williams PL, Bannister LK, Berry MM, Collins P, Dyson M, Dussek JE, Ferguson MWJ. Grays Anatomy. 38th Ed., Edinburgh, Churchill Livingstone. 2000: 1891-92.
2. Gangbo E, Lancombe D, Alberti EM *el al*. Trisomy 22 with Thyroid Isthmus agenesis and absent gall bladder. *Genet Couns* 2004;15: 311-5.
3. Sadler TW. Head and neck. *Langman's Med Embryol* 2006; 10: 270.

4. Sgalitzer KE. Contribution to the study of the morphogenesis of the thyroid gland. *J Anat* 1941; 75: 389-405.
5. Pastor Vazquez JF, Gil Verona JA, De Paz Fernandez FJ, Barbosa Cachorro M. Agenesis of the thyroid isthmus. *Eur J Anatomy* 2006; 10: 83-4.
6. Allan FD. An accessory or superficial inferior thyroid artery in a full term infant Anatomy. *Rec* 1952; 112: 53.
7. Anson BJ. Morris Human Anatomy 12th edition. The Blackinston division Mc Graw Hill Boock Co - Newyork. 1996: 168-9.
8. Marshal CF. variations in the form of the thyroid gland in man. *J Anat Physiol* 1985; 29: 234.
9. Eisler P, Der M. Levator glandulae thyreoidea und verwandte praelaryngeale Muskelbildungen. *Aant Anz* 1922: 17: 183-96.
10. Pastur VJF, De paz Fernandez FJ, Cachorro MB. Agenesis of the thyroid isthmus. *Eur J Anat* 2006; 10: 83-4.

Mantle cell lymphoma of orbit in a middle aged female-a case report

R Kumar

Department of Pathology, Universal College of Medical Sciences, Paklihawa Campus, P.O. Box 53, Bhairahwa, Nepal

Corresponding author: Dr. Rajendra Kumar, Department of Pathology, Universal College of Medical Sciences, Paklihawa Campus, P.O. Box 53, Bhairahwa, Nepal; e-mail: neerurkumar@rediffmail.com

ABSTRACT

A 31 years female presented with painful swelling, proptosis of right eyeball and headache of 9 months duration. There were absense of perception of light, pupillary reaction and extraocular movements. Exposure keratopathy with proptosis was observed. She was provisionally diagnosed a case of orbital cellulitis. The computerized tomography revealed lymphoma. Her right orbit was exenterated. The histopathologic examination revealed non-Hodgkin's lymphoma of diffuse small type. Immunohistochemistry demonstrated malignant cells of B cell origin and it was classified as mantle cell lymphoma. Orbital lymphoid tumors typically affect older persons, but occasionally, it can involve middle aged adults also. Mucosa associated lymphoid tumors are the most common affecting eye and orbit, but mantle cell lymphoma is extremely rare.

Keywords: non-Hodgkin's lymphoma, mantle cell lymphoma, diffuse small B cell lymphoma, orbit.

Orbital lymphomas are infrequent manifestation among ocular neoplasm. Lymphoma as such is a systemic disease and various types of lymphoma can involve it.[1] Non-Hodgkin's lymphomas (NHL) of orbit have been reported in the literatures which is less than 1.0% of all NHL. Although NHL of orbit is mostly indolent in nature, uncommonly it can transform into aggressive form to metastasize directly to affect eye-adnexa manifesting from swelling, compression, diplopia, ptosis, exophthalmus, sometimes enophthalmus[2] to loss of vision. Early diagnosis and proper management can save the vision and debility of the patients. Orbital lymphoid tumors typically affect older persons, but rarely, it can involve younger adults also. The latest classification of WHO divides non-Hodgkin lymphoma into B or T cell type and almost all orbital lymphomas consist of B cell tumors.[3] Mucosa associated lymphoid tumor (MALT) has been reported as the commonest tumor affecting eye and orbit, but mantle cell lymphoma is extremely rare in a middle aged person and thus it is being reported.

CASE REPORT

A 31 years female patient presented with complaints of painful swelling, proptosis of right eyeball and headache of 9 months duration. The complaints aggravated for the last 2 months followed by loss of vision. On examination, there was no perception of light, absent pupillary reaction, no extraocular movement and exposure keratopathy owing to marked degree of proptosis. The left eye was found to be normal. There was swelling and tenderness over frontal sinus. Mild lymphadenopathy of the right axilla and anemia were detected. She was provisionally diagnosed a case of orbital cellulitis, and treated with steroids and antibiotics. The symptoms relieved to some extent. The total leucocyte count was 12,500 with lymphocytosis. The computerized tomography (CT) revealed lymphoma, having a diffuse enhancing soft tissue mass within muscle cone of the right orbit replacing retro orbital fat and pushing the eyeball laterally and forward causing exophthalmos with evidence of no bony erosion. The left orbit appeared intact (Fig. 1). After intradepartmental discussion, her right orbit was exenterated under general anaesthesia and tissues were sent for histopathology examination. The tumor mass was glandular in consistency. The hematoxylin and eosin stained histopathologic examination revealed predominantly small cells, with intermediate lymphocytes in diffuse pattern having irregular nucleus and inconspicuous nucleoli. Fibrous tissue proliferation and orbital fatty tissue infiltration by neoplastic cells were seen. It was

Fig. 1. The CT scan showing diffuse enhancing soft tissue mass in the right orbit replacing retro orbital fat.

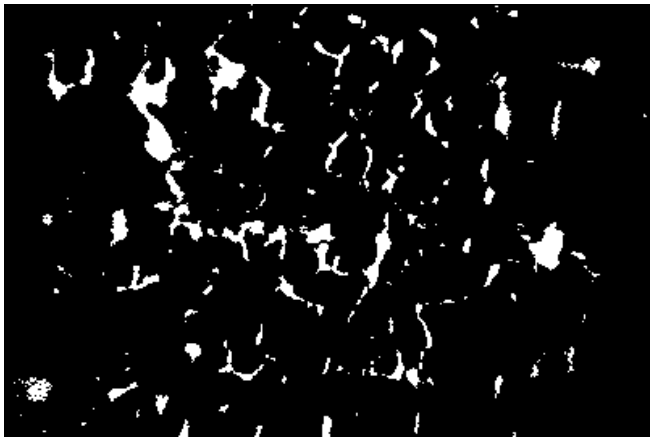

Fig. 2. Photomicrograph showing small lymphocytes in diffuse pattern (H & E x 40).

concluded with NHL of diffuse small type (Fig. 2). Immunohistochemistry demonstrated malignant clonal cells of B cell origin (CD19, CD20, CD5 positive) coexpressing surface IgM and IgD and chromosomal translocation of t (11;14). Finally it was classified as mantle cell lymphoma. CT scan of the chest and abdomen did not reveal any evidence of lymphomatous involvement. The bone marrow biopsy showed reactive lymphoid cells. The patient with preliminary advice was referred for radiotherapy for any residual involvements. She is doing well without any remarkable complaints for the last 2½ years.

DISCUSSION

The incidence of primary ocular lymphoma is although rare, it affects various parts of eye. The present case was a middle aged female. Lymphomas usually affect older age group persons above 60 years as it was reported in a 63 years female[1]. However, sometimes it has been reported in below 60 years of age. Lutz *et al* reported it in a case of 47 years male.[4] Inflammatory lesions are the most common to involve the orbit. Pseudotumour and lymphomas are next in incidence. Ptosis may result due to excessive weight of edema fluid on upper lid which creates difficulty in diagnosis as was seen in the present case with painful swelling and tender frontal sinus. In the early stage the present case was provisionally differentiated as orbital cellulitis owing to its presentation,[5] and initially prednisolone and antibiotics were advised in the case as also done in a 61 years female with complaints of headache and eyeball pain.[6] Lymphoma most commonly involves conjunctiva followed by eyelids and orbit.[7] Mantle cell lymphoma was reported involving conjunctiva in a 78 years male.[8] This lymphoma demonstrates diffuse growth pattern on histology. The microscopy of the present case showed the same predominantly of small lymphocytes with few intermediate cells. This was demonstrated histologically in 95.0% cases in a study.[9] Mantle cell lymphoma is an aggressive tumor, but MALT has been implicated as the most common tumor which involves eye and adnexa.[7] Chemotherapy and radiotherapy are vital for survival depending upon spread of disease. The role of radiotherapy in localized lesions is satisfactory and regresses the orbital signs.[4,5,9] However, in some cases, recurrence has been reported.[6] The extremely rare type of mantle cell lymphoma can affect a middle aged patient, but early presentation and investigations for diagnosis is mandatory to direct the managements.

REFERENCES

1. Yayan J, Mallouhi A, Moser P *et al*. Non-Hodgkin lymphoma of the lacrimal gland. *Ophthalmol* 2008; 105: 852-5.
2. Beden U, Sonmez B, Kandemir B *et al*. Bilateral simultaneous primary orbital lymphoma presenting with unilateral enophthalmos. *Ophthal Plast Reconstr Surg* 2008; 24: 145-7.
3. Yap YC, Ardeshna K, Kosmin A *et al*. Follicular lymphoma involving the orbit. *Ann Ophthalmol* 2007; 39: 163-5.
4. Lutz SC, Anderson SF, Wu CY *et al*. Non-Hodgkin's orbital lymphoma. *Optom Vis Sci* 2001; 78: 639-45.
5. Krist P, Plesnik J. Malignant lymphoma of the orbit. *Cesk Slov Oftalmol* 2002; 58: 247, 250-3.
6. Lee DS, Woo KI, Chang HR. T-cell lymphoma presenting as painful ophthalmoplegia. *Korean J Ophthalmol* 2006; 20: 192-4.
7. Oh DE, Kim YD. Lymphoproliferative diseases of the ocular adnexa in Korea. *Arch Ophthalmol* 2007; 125: 1668-73.
8. Miltiadis A, Spiridon G, Ioannis A *et al*. Primary mantle cell lymphoma of the conjunctiva- a case report. *Virchows Archiv* 2006; 449: 472-5.
9. Rasmussen P, Sjo LD, Prause JU *et al*. Mantle cell lymphoma in the orbital and adnexal region *Brit J Ophthalmol* 2009; 93: 1047-51.

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R Kumar

Department of Pathology, Universal College of Medical Sciences, Paklihawa Campus, P.O. Box 53, Bhairahwa, Nepal

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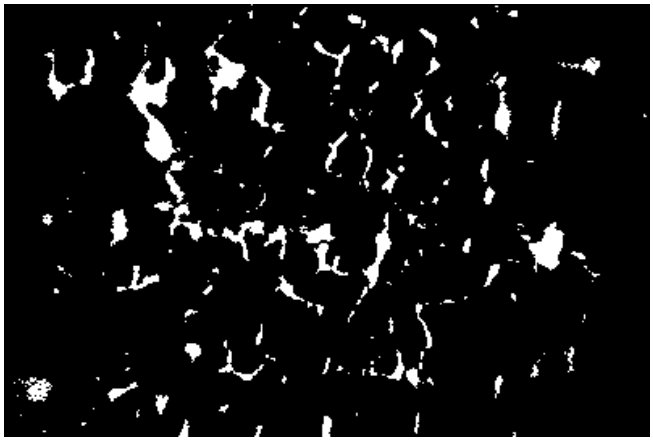

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