

TORNWALDT'S CYST: AN ABNORMAL PERSISTENT REMNANT OF NOTOCHORD – A RADIOLOGICAL STUDY WITH EMBRYOLOGICAL CORRELATION

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ABSTRACT: BACKGROUND: Tornwaldt's cyst is a midline cyst in the roof of the nasopharynx deep to pharyngobasilar fascia. It is an abnormal persistence of embryological remnant of Notochord. It causes clinical symptoms like postnasal dripping, headache and halitosis. **AIM:** To find the incidence of Tornwaldt's cyst. **MATERIALS AND METHODS:** All patients referred for MRI brain during 6 months were taken as study group. Out of 386 patients, 210 were males and 176 were females (Age: 5-86 years). In MRI, if Tornwaldt's cyst was found, its dimensions were measured and volume was calculated by the formula $l \times b \times h / 2$. **RESULTS:** Out of 386 patients, Tornwaldt's cyst was found in 3 patients (0.78%). Two patients were male and one was female. Their age ranged between 30-50 years. MRI showed the cyst was hyperintense on T2 weighted image and FLAIR in all the 3 patients. In T1 weighted image, it was isointense in 2 patients and hypointense in one. **CONCLUSION:** Despite of its rarity, Tornwaldt's cyst can mimic various pathological conditions during imaging. Therefore, its knowledge and differential diagnosis are mandatory to reach an accurate diagnosis for effective management.

INTRODUCTION: Notochord is a midline structure extending from primitive streak to prechordal plate and serves as an axial skeleton for the developing embryo¹. It is present only during intrauterine period, but disappears and turns out as remnant after birth. Some animals have persistence of entire notochord even in adult life (Example: Amphioxus)². In human beings, notochord disappears except its part which normally persists as nucleus pulposus^{1, 2} in the intervertebral discs and apical ligament of dens³.

Occasionally, abnormal persistence of remnant of notochord lead to the formation of a potential space in the postero-superior angle of the nasopharynx called Tornwaldt's bursa⁴ or Pharyngeal bursa of Luska³. Infection of the bursa obliterates its opening resulting in a cyst called Tornwaldt's cyst⁴⁻⁶.

Because of its rarity and clinical significance, the study was conducted with the aim to find the incidence of Tornwaldt's cyst and to correlate with its embryological explanations and differential diagnosis behind it.

MATERIALS AND METHODS: All patients referred for MRI study of brain in the span of 6 months duration were taken as the study group. Out of 386 patients, 210 were males and 176 were females. Their age ranged between 5 to 86 years. MRI scan was done using 1 Tesla SIEMENS MRI scanner (closed type). Head coil were used for getting good resolution images. Routine sequences of Axial TSE T1, TSE T2, FLAIR, DWI & ADC, Sagittal TSE T1 and Coronal TSE T2 were taken. Routine MR imaging of brain in axial, sagittal and coronal planes normally includes the posterosuperior aspect of

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nasopharynx also. The images were analyzed with the special attention on the posterosuperior aspect of nasopharynx. If Tornwaldt's cyst was observed, its dimension in craniocaudal, anteroposterior and transverse diameter were measured and the volume was calculated by using the ellipsoid formula $(l \times b \times h) / 2$.

RESULTS: Out of 386 patients, Tornwaldt's cyst was found in 3 patients (0.78%). Out of 3 patients, two were males and one was female. The age of the patients having Tornwaldt's cyst was between 35-50 years.

In two patients, Tornwaldt's cyst was symptomatic and associated with the nose and throat symptoms. Headache was present in those two patients; foreign body sensation in one of those patients and halitosis in another patient. However for the third patient, the cyst was asymptomatic. The patient was referred for blurring of vision. MRI findings showed normal brain and orbit and the Tornwaldt's cyst was diagnosed incidentally.

The shape of the Tornwaldt's cyst in MRI was oval in 2 patients and round in 1 patient. The cyst had characteristic signal intensity in MRI. In T2 weighted and FLAIR images, the cyst was hyperintense in all the 3 patients (Fig 1b, 1c, 2b, 2c, 3b, 3c); whereas in T1 weighted image, the cyst was isointense with muscles in two patients (Fig 1a, 2a) and hypointense in 1 patient (Fig 3a).

The measurement of maximum and minimum mean diameter of the cyst in this study was 7.47 mm and 9.73 mm respectively. The calculated volume of the cyst ranged between 0.2 to 0.4 cc.

The details of the patients having Tornwaldt's cyst in this study are tabulated below (Table 1):

	Age	Sex	Symptoms	Diameter of the cyst (mm)			Mean size of the cyst	Volume
				Vertical	Antero-posterior	Transverse		
Case 1	48	Male	Headache & halitosis	8.3	7.2	6.9	7.47	0.2
Case 2	35	Male	Headache & foreign body sensation	12.5	10.6	6.1	9.73	0.4
Case 3	50	Female	Blurring of vision	10.2	7.4	5.3	7.63	0.2

Table 1

DISCUSSION:

HISTORY: In 1840, Mayor was the first person to find the Tornwaldt's cyst in an autopsy case⁴. However, in 1885, German physician Gustav Ludwig Tornwaldt reported 26 cases of Tornwaldt's cyst, considering it as pathological and hence the cyst was named after him^{4,7,8}. In 1912, Huber gave an embryological explanation that this is a space where the notochord has retained its union with the pharyngeal endoderm^{4,6,9}.

Embryology:

During third week of development, pre-notochordal cells invaginating inside the primitive pit move forward and cephalad upto prechordal plate. These pre-notochordal cells intercalate with underlying hypoblast so that, for a short time a double layered plate called notochordal plate is found in the midline of the embryo. When the hypoblast cells are replaced by the cells of endoderm moving in at streak, notochordal cells detach from endoderm to form the definitive notochord¹.

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When the notochordal cells intercalate with pharyngeal endoderm, a small out pouching develops from pharyngeal mucosa called Sessel's pouch, thereby a focal adhesion is created between notochord and endoderm. During sixth week of development, when the most cephalic portion of notochord regresses, its remnant may persist along with its out pouching in the pharyngeal endoderm⁶. This communication may provide a pathway forin-growth of respiratory epithelium, forming a potential space leading to formation of Tornwaldt's bursa. Due to repeated infection and inflammation, fluid collects in the space and its mouth is obliterated creating a cyst called Tornwaldt's cyst⁴⁻⁶.

A unique case was reported recently¹⁰ about the association of Tornwaldt's cyst with cerebral artery abnormalities and infarction. It was suggested that this association may be due to dysfunction in cell signaling via Notochord-derived hedgehog (Hh), a key molecular for angiogenesis to form appropriate blood island in yolk sac.

INCIDENCE: Incidence of Tornwaldt's cyst in autopsy studies was 1.5% to 3.3%^{4,11}. However, as per MRI reports its incidence was 0.2% to 5%¹². In 1998, Lkushima et al⁶ reported that its incidence in MRI was 1.9%. In 2007, Moddy et al⁴ reported that its overall incidence was 0.06% (32 out of 52, 013 cases) of which incidence in CT was 0.013% and by MRI was 0.13%. The last case of Tornwaldt's cyst case was reported in 2013 by Brenten PI et al¹³. The incidence of the present study was 3 out of 386 patients (0.78%).

The incidence in the studies conducted recently was found to be lesser than older studies, showing the rarity of Tornwaldt's cyst in recent days. This may be probably due to prenatal usage of high dose of folic acid nowadays⁴.

Clinical presentation:Tornwaldt's cyst can occur in any age group and there is no sex predisposition. However, its peak incidence was noticed in the age group of 15 to 30 years⁵. Contrary to this, Lkushima et al⁶ observed its incidence was high in the age group of 40 to 60 years. In the present study, the age of the patients was between 30-50 years.

The size of Tornwaldt's cyst is variable. The smallest and largest measurements reported so far reported are 2 mm¹⁴ and 25 mm⁷ respectively. According to the study conducted by Moddy et al⁴, the mean size of Tornwaldt's cyst as per CT & MRI findings was 0.66 cm³ and 0.58 cm³ respectively. In the present study, maximum and minimum mean diameter of the cyst measured 7.47 mm and 9.73 mm respectively. The volume of the cyst ranged between 0.2 to 0.4 cc.

Tornwaldt's cyst is of two types. If the cyst drains into nasopharynx, it is called crusting type. If it does not drain, it is called cystic type⁴. In the present study, the cysts were not draining into nasopharynx. So they are of cystic variety.

In few studies, it was reported that more than half of the patients with Tornwaldt's cyst had a previous history of tonsillectomy and adenoidectomy¹⁵. It was suggested that traumatic manipulation of nasopharynx in the form of tamponade insertion, trauma and surgery like adenoidectomy may predispose to Tornwaldt's cyst⁶. Controversy to this, in the study conducted by James et al⁵, none of the patients had previous history of surgery in the area of nasopharynx. In the present study also, there was no history of trauma or surgery which is in concordance with James et al⁵ study.

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Initially Tornwaldt's cyst is asymptomatic. After infection, it is associated with group of symptoms referred as Tornwaldt's syndrome. It includes persistent posterior nasopharyngeal drainage, dull occipital headache, and stiffness of the posterior cervical muscles, increased pain associated with movement of the head, halitosis and an unpleasant taste¹³ and even habitual snoring¹⁷. In the present study, Tornwaldt's cyst was symptomatic in two patients and asymptomatic in one patient.

Apart from these symptoms, it was reported that Tornwaldt's cyst may be associated with uncommon complications like isolated sphenoidal pyocele¹⁸, cerebral arteritis and stroke¹⁰.

Radiological findings: The signal intensity of the lesion on T1 weighted images may vary from intermediate to high, depending on the protein content or hemorrhage in the cyst, whereas on T2 weighted image, usually found to be high⁸. In the present study also, in T2 weighted image and FLAIR image, the cyst was hyperintense on in all the three patients. Whereas, in T1 weighted image, it was isointense in two cases and hypointense in one case.

Differential diagnosis: In the present cases, Tornwaldt's cyst was located deep to pharyngobasilar fascia. Thereby, the chance of being a 'retention cyst of pharyngeal tonsil' was excluded, because retention cyst usually lies superficial to pharyngobasilar fascia. The difference between the Tornwaldt's cyst and retention cyst can be made out by the histological fact that retention cyst shows presence of lymphatic follicle¹⁹.

In the present cases, the cyst had not eroded the bony structures or sphenoid air sinus. There was no thickening of soft tissue in nasopharynx. Therefore, the possibility to be an inflammatory mass or nasopharyngeal neoplasia was excluded^{5, 20}. Meningocele, meningoencephalocoele, sphenoid sinus mucocoele are usually associated with bony defects⁶. These cyst can neither be a branchial cyst from second pharyngeal cleft nor be a Rathke's pouch. These two embryological remnants can be differentiated by the position. Branchial cyst from second pharyngeal cleft are found in the region of Eustachian cushion, lateral to the midline. But in the present cases, the cysts were exactly in the midline. Rathke's pouch is located more ventral and cephalad to the notochord remnant^{5, 6}.

The cysts in the present cases were least likely to be mucous glands cyst, because mucous gland cysts are usually multiple, smaller in size, occur outside the mid-line and contain more lymphoid tissue than Tornwaldt's cysts⁵.

Adenoid retention cysts are multiple and of size less than 5 mm. In the present cases, the cysts were single and the size was more than 5 mm.

Some of the benign vascular lesions of nasopharynx which mimic Tornwaldt's cyst are Juvenile angiofibroma, hemangioma and hemangiopericytoma. The correct diagnosis of Tornwaldt's cyst helps in proper management. Simple drainage of the cyst will lead to recurrent episodes due to reinfection and fluid retention. The treatment of choice is surgical removal or marsupialization⁵. Transnasal endoscopic marsupialization provides excellent surgical field of vision to avoid damaging the Eustachian tube. The root of the cyst may be rooted to underlying prevertebral fascia and complete exenteration of the cyst would be best done under general anesthesia⁸.

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CONCLUSION:In the present study, the incidence of Tornwaldt's cyst in this study was 0.78%. Two patients were male and one was female. The age of the affected patients was between 35-50 years. In T1 weighted image, the cyst was homogenously isointense in 2 patients and hypointense in 1 patient. In T2 weighted image and FLAIR image, the cyst was hyperintense in all the 3 patients.

Therefore, it is concluded that when a patient with the complaints of postnasal drip, occipital headache and halitosis is presenting with radiographic appearance of soft tissue mass projecting in the postero-superior aspect of nasopharyngeal space without any bony erosion and soft tissue reaction, the diagnosis of Tornwaldt's cyst should be considered. Prompt surgical intervention at right time may alleviate all the symptoms.

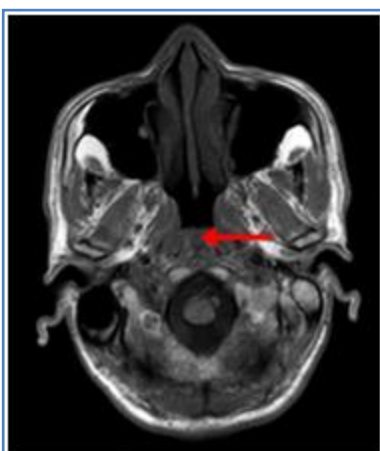


Fig. 1a
Axial T1 weighted image

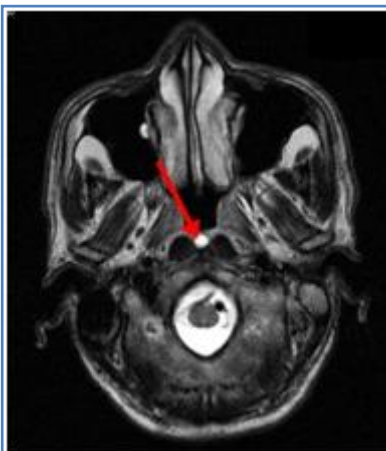


Fig. 1b
Axial T2 weighted image

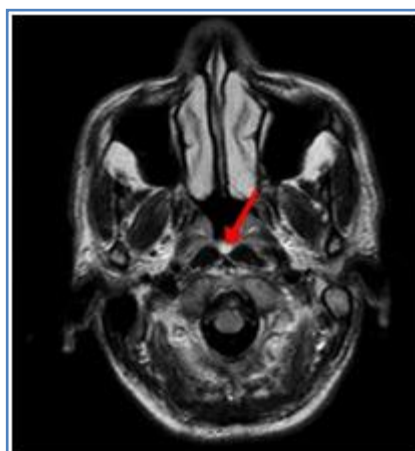


Fig. 1c
Axial - Flair

Fig. 1a, 1b, 1c:Axial MRI of Case-1 shows a well circumscribed thin walled round cystic lesion in the midline of roof of nasopharynx. It is homogenously isointense on T1 weighted image (Fig 1a) and hyperintense on T2 weighted image and FLAIR (Fig 1b & 1c). It is in the submucosal plane between the longus colli muscles without any inflammatory changes.



Fig. 2a
Sagittal T1 weighted image



Fig. 2b
Coronal T2 weighted image

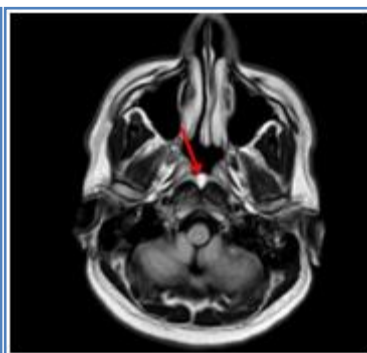


Fig. 2c
Axial - Flair

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Fig. 2a, 2b, 2c: MRI images of Case-2 shows a well-defined oval shaped thin walled cystic lesion in submucosal plane in the roof of nasopharynx. It is homogenously isointense on Sagittal T1 weighted image (Fig 2a) and hyperintense on Coronal T2 weighted image (Fig 2b) as well as in Axial FLAIR image (Fig 2b & 2c).

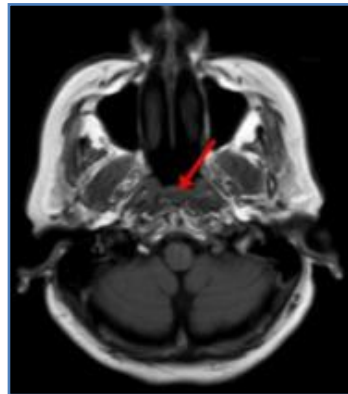


Fig. 3a
Axial T1 weighted image

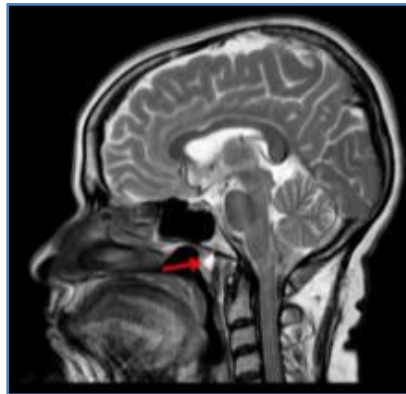


Fig. 3b
Sagittal T2 weighted image

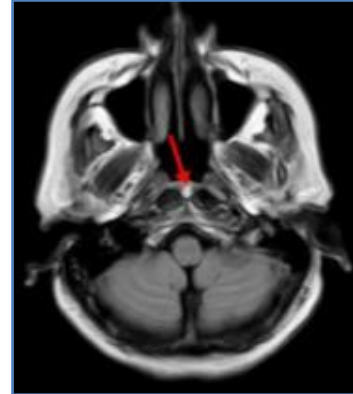


Fig. 3c
Axial - Flair

Fig. 3a, 3b, 3c: MRI of Case-3 shows a well-defined oval shaped cystic lesion in the midline of roof of nasopharynx. It is homogenously hypointense on Axial T1 weighted image (Fig 3a), hyperintense on Sagittal T2 weighted image (Fig 3b) and Axial FLAIR image (Fig 3c). It is in the submucosal plane between the longus colli muscles. No septations or solid components are seen.

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