Immature Teratoma of Head and Neck: Two case reports

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Teratoma refers to a neoplasm that recapitulates all three germ layers. Teratoma is usually developmental and sometimes congenital neoplasm which displays both solid and cystic components with gross and microscopic differentiation into a wide variety of tissues representative of all three germ layers i.e. ectoderm, mesoderm and endoderm. Teratomas of the head and neck are extremely rare and usually seen during the neonatal period. Teratomas of the head and neck due to their obscure origin, bizarre microscopic appearance, unpredictable behavior and often dramatic clinical presentation are a clinical surprise. We are presenting two case reports, where immature teratoma was present in hypopharynx and in nose.

Key words: Immature teratoma, nose, Hypopharynx, totipotent, germ layers

INTRODUCTION

Teratomas are embryonal neoplasms that arise when totipotential germ cells recapitulate normal organogenesis and give rise to more or less organoid masses in which tissues derived from all three blastodermic layers (ectoderm, endoderm, and mesoderm) can be identified (Chakravarti, 2011; Rothschild, 1994). Broadly, these are classified as mature, immature and malignant. Age range at presentation varies from birth to 6th decade of life (Ling, 1993). The immature form of teratoma was first described in 1960 by Thürlbeck and Scully (Trabelsi, 2002). It is found either in pure form or as a component of a mixed germ cell tumor. According to WHO, immature teratoma (IT) is defined as a teratoma containing a variable amount of immature embryonal type (generally) neuroectodermal tissue.

They occur in genital systems and other organs along the midline of the body, with similar morphology and classification (Azizkhan, 1996). The incidence of teratomas is 1 per 4000 live births, with the sacrococcygeal area being the most common site (Coppit 2000). Teratomas of the head and neck are extremely rare and teratomas occur in these locations in nearly 1-3.5% of all cases (Dadmehr, 2006). The common sites of involvement are the neck, oropharynx, nasopharynx, orbit, and paranasal sinuses. Most neck teratomas are reported as mature, and malignant immature teratomas have been said to occur in approximately five percent of teratomas of the neck. The current treatment for tumor is surgical excision with adjuvant chemotherapy. Cisplatin, etoposide, and bleomycin (BEP) are commonly using chemotherapeutic agents for immature teratoma.

We are presenting here two case reports. In case one it involved hypopharynx and in second case it involved nose.

Case report 1

Fifty five years male patient was presented with history of difficulty in swallowing for solid more than liquid for the last 6 weeks.

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He also had history of change in voice and difficulty in breathing for the last two weeks. He presented with stridor in our OPD and indirect laryngoscopic examination showed single globular mass present in right side of hypopharynx and covering endolarynx completely. Neck examination was unremarkable. The rest of local examination was normal. In view of stridor, emergency tracheostomy was performed. The X ray soft tissue neck lateral view showed mass in hypopharynx covering larynx (Figure 1).

Direct laryngoscopy revealed a single globular pinkish white mass with irregular surface (4×4 cm in size) which was found attached with right lateral wall of pyriform sinus by narrow stalk (Figure 2).

Endolarynx and left half of hypopharynx was normal. The base of the mass was cauterized and excised completely. The patient was kept on nasogastic tube for five days. Tracheostomy tube was removed on 6th postoperative day. The histopathological examination showed mature pilosebaceous units along with immature mesenchymal elements revealed and diagnosis of immature teratoma was made (Figure 3). After second cycle of chemotherapy, the patient expired.

Case report 2

Twenty year male patient presented with history of left side nasal bleeding for the last one month. The bleeding was spontaneous, episodic and profuse in amount. He also complained of nasal obstruction and loss of smell from left side for the same duration. There was no history of headache, nasal discharge, facial swelling, decreased vision and diplopia. Patient was referred to our department with biopsy report of angiomatous polyp in left nasal cavity. On examination, reddish fleshy mass was coming out from left nasal cavity with widening of nasal ala (Figure 4). Nasal septum was deviated to right side. The rest of local examination was normal. Contrast enhancing CT scan of nose and paranasal sinuses showed homogenous soft tissue density in midline involving left side nasal cavity, ethmoid sinuses, nasopharynx with retained secretion in left maxillary sinus and frontal sinus (Figure 5). Cribriform plate was seemed to be eroded by disease. Keeping the possibility of esthesioneuroblastoma as diagnosis, the mass protruded from left nasal cavity was removed with cautery. As there was no bleeding, the tumor was completely removed endoscopically. The tumor was soft and friable and extended posteriorly till choana with attachment on the posterior part of septum. The final biopsy report showed focally hyperplastic respiratory epithelium lined tissue...
Figure 4. Radish fleshy mass coming out from left nasal cavity extending till upper lip with widening of nasal ala.

Figure 5. Contrast enhancing CT scan of Nose and PNS (coronal cut) showing homogenous soft tissue density in left side of nasal cavity and paranasal sinuses with erosion of septum and cribriform plate.

Figure 6. Photomicrograph showing focally hyperplastic respiratory epithelium lined tissue showing immature mesenchymal elements in the subepithelium (HEX200).

showing immature mesenchymal elements in the subepithelium, which was suggestive of immature teratoma (Figure 6). In post operative period, the patient received chemotherapy and has been regular follow up for last 1 year with no recurrence.
DISCUSSION

Teratomas are common tumors originating from totipotential cells and are composed of heterogeneous tissues reminiscent of derivatives from any or all three germ layers in varying ratio (Duwe, 2005). The presence of tissues within the lesion that are foreign to the affected sites is a distinctive feature of this rare tumor. Most of the cases occur in neonates and older infants, contrary to our patient who were adult age-group. It has been established that there is no sex predilection in head and neck teratomas (Abemayor, 1984). A mature teratoma is typically benign and is found more commonly in females, but immature teratomas are typically malignant and are found more often in males as in our cases. As a general rule, while pediatric teratomas of the head and neck tend to be oncologically benign, adult teratomas tend to be histologically and oncologically malignant. The duration of symptoms is short in malignant form as compared to benign form of immature teratoma was seen in our cases (Ibrahim, 2012; Swetha, 2011).

Benign teratomas are solid and cystic tumors composed of a variety of both immature and mature tissues derived from all three germ layers. The epithelial component of a benign teratoma usually consists of mature squamous epithelium and immature intestinal or respiratory epithelium. Primitive neuroepithelium with rosettes, pseudorosettes or neurofibrillary matrix predominates in some tumors. Pigmented retinal epithelium can also be seen. The mesodermal component consists of fibroblasts and embryonic, immature spindle cells embedded in a myxoid matrix. Islands of cartilage, smooth muscle cells, and skeletal muscle cells exhibiting varying degrees of maturity may also be present. To make the diagnosis of teratoma, it is mandatory to find at least two of three germ layers (Smirniotopoulos, 1995). Teratomas may also be histologically immature while being oncologically benign, or they may harbor malignant components and have the potential to exhibit an aggressive biological behavior. The amount of neuroectodermal immature tissue present permits the classification of immature teratomas into three grades of increasing malignancy. Immature teratoma may metastasise to brain, liver and lung. Metastasis to brain has also been reported. The imaging appearance is typically of a large, heterogeneous mass with a prominent solid component. However, the spectrum of appearances ranges from a predominantly cystic to a predominantly solid mass. The presence of a prominent solid component containing calcifications and small foci of fat. Cystic components may contain serous, mucinous, or fatty sebaceous material.

The recommended management for head and neck teratomas is complete surgical excision. Chemotherapy has been successful in those with recurrent disease as this tumor is highly chemosensitive (Ernest E. Lack 1985). In both cases we followed the same treatment protocol as per literature. The prognosis with tumor remains guarded, it depends on early detection and aggressive management. We learn from these two cases that immature teratoma can involve any part in head and neck, it require early diagnosis and management.

REFERENCES


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