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Histopathology

PRIMARY TERATOMAS OF UNUSUAL LOCATIONS: A CASE SERIES AND A REVIEW OF LITERATURE

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ABSTRACT Teratomas are common germ cell tumours made up of tissues from the germ cell layers of ectoderm, endoderm, and mesoderm. The ovaries and testes are the most typical sites of occurrence. However, teratomas can be observed in various anatomical locations. Head and neck teratomas are rare benign tumours, and in children they are usually diagnosed early due to the frequent symptoms. Exceptionally rare cases are found in the thyroid gland. Extragonadal abdominopelvic teratomas are rare, with the spleen being an unusual site. They are often congenital and rarely turn malignant. We report a series of three cases of mature teratoma in unusual sites of the thyroid, cheek, and spleen. **Summary:** Teratomas are tumours that originate from germ cells and are more common in the gonads, sacrococcygeal region, and mediastinum. They can also occur in the head and neck regions, extragonadal abdominopelvic regions, and other rare sites. They are often congenital and rarely turn malignant. Since teratoma-specific radiological features are not clear and clinical characteristics are still not well characterised, surgical resection and histological examination are required for diagnosis.

KEYWORDS : Teratomas, Thyroid, Mature, Spleen, Extragonadal

INTRODUCTION

Teratomas are tumours that originate from germ cells and are made up of tissues from the ectoderm, endoderm, and mesoderm germ cell layers. They are more common in the gonads, sacrococcygeal region, mediastinum, and pineal region. They can also occur in the head and neck regions, predominantly in the cervical, nasopharynx, face, and orbital regions. Exceptionally rare cases are found in association with the thyroid gland [1]. Of all teratomas, 3-6% are encountered in the head and neck regions [2]. In the literature, the cheek is the least frequently mentioned site of teratoma [3-5]. Extragonadal abdominopelvic teratomas are rare, representing 1-5% of germ cell tumours. Among them, splenic teratomas are extremely uncommon, with very few case reports in the literature [6]. It is difficult to specify their clinical features in these uncommon locations due to their rarity. Herein, we report a series of three benign teratoma cases in unusual locations like the thyroid, cheek, and spleen.

CASE REPORT

We report three cases of teratomas.

Case 1

A 7-year-old boy had a cervical mass that had increased in size for the past year. There were no associated local or systemic symptoms. Ultrasonography of the neck showed a large mixed echogenic lesion with solid, cystic, and calcific components in the left paramedian area in the region of the left lobe of the thyroid. A thin rim of the left lobe was seen. The lesion measured 6*3.5*2.3cm. Minimal vascularity was noted. The lesion was seen displacing the left carotid and jugular vessels laterally (Figure 1A). The possibility of left lobe thyroid nodule TIRADS 4 was suggested and advised histopathological correlation. The left lobe of the thyroid gland was sent for a frozen section. The lobe of the thyroid gland measured 6.5*4*3.2cm. The external surface appeared congested. Serial sectioning revealed a well-circumscribed nodule with a solid-cystic cut surface measuring 6.2*4.5*3cm. Solid areas were greyish-white and myxoid. Cystic areas were filled with mucin. The lesion extended up to the inked outer surface and involved the isthmic margin. The frozen section was reported as lesional tissue composed of neural elements (Schwannian stroma) with extensive psammomatous calcification and a focal mucous lining with the periphery of a normal thyroid gland. Lesional tissue was present at the margin.

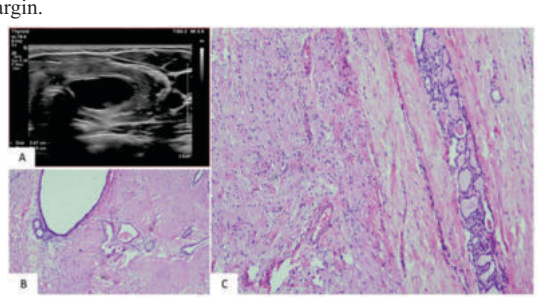

Figure 1: (A) Ultrasonography of the neck reveals a large mixed echogenic lesion with solid, cystic, and calcific components. (B and C) H and E sections show diffusely proliferating neuroglial tissue, cystic ducts, a few intestinal glands with mucin, and stromal muscle with blood vessels. (B and C at 10x magnification).

Permanent sections showed diffusely proliferating neuroglial tissue arranged as vague nodules with extensive areas of psammoma bodies and patchy areas of ganglion cells; cystic ducts filled with foamy histiocytes; papillary plexus (choroid plexus); a few intestinal glands with mucin; focal lymphoid aggregates; and stromal muscle with blood vessels (Figures 1B and C). The isthmus showed compressed reactive follicles and muscle bundles, free of tumour. There was no evidence of undifferentiated areas, primitive neuroepithelial cells, or necrosis. These findings supported the diagnosis of a mature teratoma involving the left lobe of the thyroid. The patient was lost to followup.

Case 2

An 8-year-old girl presented with complaints of recurrent right cheek swelling since the age of three. Records of previous surgical events were not available. MRI of the neck revealed an ill-defined T1 and T2 hyperintense lesion seen in the right buccal and masseteric space measuring approximately 6.6*5.2 cm with fibrous bands and mesenchymal elements within (Figure 2-A). These features were suggestive of lipoblastoma in the right buccomasseteric space. The specimen was received as multiple fragments of fibrofatty tissue ranging in size from 1 cm to 6.5 cm in the greatest dimension. Serial sectioning revealed pale greyish-white areas that were variable in consistency. Peripherally attached focal skeletal muscle fibres were also noted. Histological examination showed a vaguely circumscribed lesion composed of lobules of adipocytes separated by paucicellular fibrovascular septae. The adipocytic component had mature fat displaying mild to moderate variation in size, and fibrovascular septae showed ectatic anastamosing vasculature lined by prominent endothelial cells, the choroid plexus, and smooth muscle bundles. Also seen in the septae are scattered lymphoid aggregates with psammomatoid calcifications, degenerated and atrophic skeletal muscle fibres, nerve bundles, lobules of cartilage, and minor salivary glands. No nuclear atypia, immature neuroepithelial elements, or overt features of malignancy were noted (Figures 2B and C). These findings pointed towards the diagnosis of a mature teratoma, right cheek swelling.

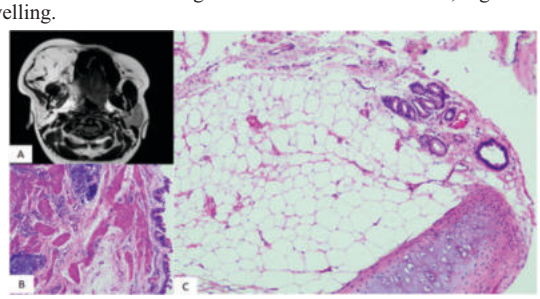

Figure 2: (A) MRI of the neck reveals an ill-defined hyperintense

lesion in the right buccal and masseteric space with fibrous bands and mesenchymal elements within. (B and C) H and E sections show bundles of smooth muscle fibres, scattered lymphoid aggregates, lobules of adipocytes, cartilage, and minor salivary glands (B and C at 10x magnification).

Case 3

An 18-year-old boy presented with complaints of abdominal pain and discomfort for the past 2 years. He had a previous history of debridement of the splenic cyst about 3 years ago, and the biopsy was reported as inconclusive. On physical examination, there were no respiratory or neurological deficits. An abdominal examination showed mild splenomegaly. Laboratory workup revealed mild neutrophilia, and the rest of the haematological and biochemical parameters were within normal limits. A CT scan of the total abdomen revealed an enlarged spleen with a large, thin-walled, multiloculated, complex, non-enhancing cyst in the superior polar parenchyma with hilar and subcapsular extension. A few tiny foci of discontinuous wall calcifications were noted (Figure 3-A). Following these investigations, an open splenectomy was performed. As the cyst was large with subcapsular extension, splenectomy was a matter of choice.

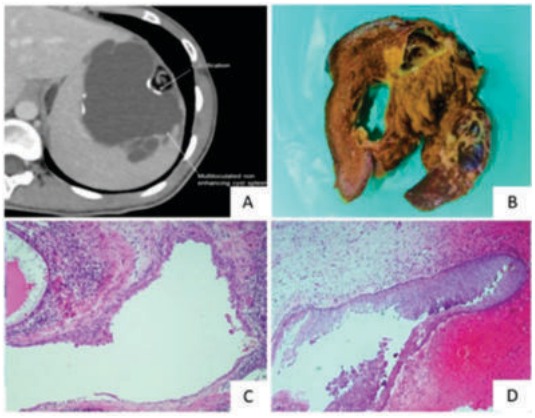

Figure 3: (A) A CT image reveals an enlarged spleen with a large, thin-walled, non-enhancing cyst with subcapsular extension and foci of wall calcification. (B) Gross image of the spleen with multiloculated, thin-walled cysts filled with fibrofatty soft tissue. (C and D) Hand E sections show cysts lined by a single layer of cells and also by squamous epithelium. (C and D at 10x magnification).

Grossly, we received an enlarged spleen measuring 14 cm in the greatest dimension and weighing 463 g. The capsular surface showed a greyish-tan ragged area. Serial sectioning revealed a multiloculated, thin-walled cyst replacing the splenic parenchyma, measuring 9.5 x 7.7 x 5.5 cm. The cysts contained tan-brown-coloured fluid. The inner wall of the cyst was greyish white to tan yellow, firm, and glistening with intervening coarse fibrous trabeculations. Some of the cysts were filled with yellowish, fibrofatty soft tissue with foci of calcification and haemorrhage (Figure 3-B). Microscopic examination revealed multiple cysts lined by a single layer of epithelium and foci of squamous epithelium. The cysts were predominantly enclosed with cyst macrophages in hemorrhagic background. Foci of cysts composed of lobules of mature adipocytes, smooth muscle fibres, and areas of calcification were noted (Figures 3C and D). A diagnosis of mature cystic teratoma was rendered.

DISCUSSION

Teratomas are tumours that originate from germ cells and are made up of tissues from the ectoderm, endoderm, and mesoderm germ cell layers. They are more commonly encountered in the gonads, sacrococcygeal region, mediastinum, and pineal region. They can also occur in the head and neck regions, predominantly in the cervical, nasopharynx, face, and orbital regions, accounting for 3% to 6% of all teratomas [1, 2]. Since they frequently exhibit symptoms including respiratory distress, facial deformity, and orbital involvement in children, they are typically identified at an early stage. Unlike in adults, teratomas in children are typically congenital, and they very seldom become malignant. Other rare sites of teratomas described in the literature are the gluteal region, the palm, the retroperitoneal space, the floor of the mouth, the urinary bladder, the kidney, and the stomach [7-10]. In our study, we report a series of three benign teratoma cases in the thyroid, cheek, and spleen.

To diagnose thyroid teratomas, the tumour must occupy a part of the thyroid, be in direct continuity with the thyroid, or exist in the neck with the thyroid being incompletely developed [11-13]. In our case of thyroid teratoma, the tumour was almost entirely occupying the left lobe of the thyroid gland and displacing the major vessels of the neck, but without any compression of the airways. Radiologically, the presence of calcifications on the plain film or mixed echogenicity with multiloculated cystic or solid regions on ultrasonography are clues to diagnosing teratomas [14]. However, teratoma-specific ultrasonic features are not clear, and our case of cheek teratoma was radiologically diagnosed as lipoblastoma, revealing their unclear image findings. Lymphoid hyperplasia, lymphoma, cystic hygroma, thyroglossal duct cyst, and other congenital malformations should be considered in the differential diagnosis when a child presents with a neck mass [14, 15]. Based on the site and size of the lesion, teratomas of the neck can cause symptoms like airway compression and are thus detected early. In children, cervical teratomas are mostly benign, but the likelihood of malignant teratomas is higher in adults [13, 15]. Surgery must be performed to diagnose and treat malignant tumours since diagnostic imaging methods are imprecise in distinguishing them from benign tumours [14]. The goal of surgery is to completely remove the lesion while leaving the normal structures in place.

Histologically, teratomas are made up of tissues from the ectoderm, endoderm, and mesoderm germ cell layers, imparting a heterogenous histological picture. It can include solid and cystic areas with mature or immature components. In our case of thyroid teratoma, there was diffusely proliferating neuroglial tissue, patchy areas of ganglion cells, and a few intestinal glands with mucin, supporting the diagnosis of a mature teratoma. Also, the case of the cheek teratoma had lobules of cartilage, adipocytes, smooth muscle bundles, and salivary acini with no evidence of immature elements.

True cysts of the spleen encompass both parasitic and nonparasitic cysts. Parasitic cysts are the most common cause of splenic cysts, which are caused by Taenia echinococcus. Non-parasitic cysts are subclassified into congenital, vascular, and neoplastic cysts. Cysts of congenital origin include epidermoid, dermoid, and endodermoid cysts. However, the classification of splenic cysts continues to evolve, and lining alone cannot be a reliable criterion for classifying the cyst [16]. There are various hypotheses regarding the etiopathogenesis of splenic cysts. Congenital cysts have been attributed to the developmental misplacement of epithelial tissue during embryogenesis with consequent metaplasia. However, dermoid cysts originate from the invagination of the splenic capsular mesothelium during development with consequent fluid accumulation for unknown reasons, resulting in cyst formation [17]. Splenic cysts are asymptomatic in 30-60% of patients; most of them are diagnosed incidentally. Ultrasound is the preferred initial modality, which reveals whether the cyst is unilocular or multilocular, with or without solid components. CT and MRI imaging are adjuvant modalities that provide additional information. In cases of pure cystic lesions, epidermoid cysts, echinococcal cysts, hematomas, lymphangiomas, and pseudocysts are considered differential diagnoses [18]. Whereas in the case of a cyst with partly solid areas showing fat and calcifications, splenic teratomas are considered [16]. Histologically, the splenic teratoma had the same features as mature cystic teratomas in any other location.

Treatment approaches such as aspiration and sclerosis, internal and external marsupialization, partial cystectomy, and partial splenectomy have all been described. But in these treatment approaches, the rate of cyst recurrence is common. Hence, the treatment for splenic cysts is total splenectomy [19]. Recurrence in mature teratomas is uncommon and has an excellent prognosis. Our cases were lost to follow-up, causing an inability to comment on the recurrence rate of teratomas in these unusual locations.

CONCLUSION

Teratomas in the head and neck region and thyroid are rare benign tumours; however, some teratomas are malignant. The differential diagnosis of cystic lesions in the head and neck region of children should take thyroid teratomas into consideration. The cheek is one of the least frequently mentioned sites of teratoma. In cases of splenic cysts with partly solid areas and focal calcifications in their walls, a diagnosis of splenic teratoma is considered. Since teratoma-specific radiological features are not clear and clinical characteristics are still not well characterised, surgical resection and histological examination are required for diagnosis. Even odd places should not be a problem

with the characteristic histological features for teratoma diagnosis.

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