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CASE REPORT Open Access

# Brain ectopic tissue in sacrococcygeal region of a child, clinically mimicking sacrococcygeal teratoma: a case report

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## Abstract

**Background:** Mature brain heterotopic tissue in sacrococcygeal region is a very rare benign congenital abnormality of newborn. To date, only two cases of mature heterotopic brain tissue in the sacrococcygeal region is reported by literature. Heterotopic brain tissue in other areas such as lung, nose, face and retroperitoneal region are also rarely reported. Meanwhile, rather than brain heterotopic tissue in sacrococcygeal region, a case of adrenal gland heterotopic tissue in sacrococcygeal region also has been reported.

**Case presentation:** A 3.5 month-old male baby presented with history of sacrococcygeal mass since birth. Clinical examination of the child was good with no other problem. Sacrococcygeal region revealed an elevated round mass with no discharge. Computed tomography reported a large sacrococcygeal teratoma type-III arising from the sacrococcygeal region extending intra-abdominally to the level of L2 vertebral body. The mass was excised by the impression of sacrococcygeal teratoma (SCT). On gross examination, a gray-white irregular tissue fragment with 5 cm in greatest dimension was examined. Cut sections showed homogenous yellowish white appearance. Histological examination revealed solid fragments composed of mature neural tissue comprising glial cells and astrocytes with no other germ cell layer component.

**Conclusion:** Mature brain heterotopic tissue in sacrococcygeal area is a rare benign disease. Two ectopic brain tissue in sacrococcygeal region were previously also reported. Sacrococcygeal teratoma is the most common congenital tumor, but this current rare case of heterotopic brain tissue in sacrococcygeal region should also be in the differential diagnosis.

**Keywords:** Congenital malformations, Heterotopic brain tissue, Sacrococcygeal teratoma

## Background

The sacrococcygeal region is the most affected area for congenital rare abnormalities and birth defects in neonatal period. The germ cell tumors are the most common form of these anomalies (Shrestha et al. 2016; Sugathadasa et al. 2013). Congenital abnormalities are a permanent change in body structure due to intrinsic anomaly of the body structure in prenatal period. The prevalence of major congenital malformations which are reported in different population around the world is ranging from less than 1% up to 8% (Singh et al. 2014). Prevalence of congenital abnormalities can be varied in different population. Genetic and environmental factors along with poor nutrition are causes of congenital malformations in newborn (Bhandari et al. 2015). A study in UAE showed 173 babies with major congenital malformations during 2 years' period, revealing the incidence of 10.5/10000 births (Gazali et al. 1995). Congenital neoplasms caused 1.5–2% of all pediatric neoplasms with a prevalence rate of 1/12500 to 1/27500 live births (Alamo et al. 2011). The most common of these abnormalities in sacrococcygeal region is SCT. The majority (75%) of SCT are benign and rarely (12%) they are malignant and life threatening (Shrestha et al. 2016). These tumors are believed to arise from totipotent cells of Hense's node, which is remnant of the primitive streak in sacrococcygeal area in early gestation (Mandal

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et al. 2015). Complete resection of the tumor for most patients have good outcome, but some can develop high-out-put cardiac failure and hydrops because of arteriovenous shunting (Langer 1993). Unusual course and multiple distant recurrence of SCT is also reported (Bechtel and Gauger 2014). Sacrococcygeal region neoplasms and anomalies other than SCTs are: myelomeningocele, myxopapillary ependymoma, fetus in fetu, Curarinos syndrome, ano-rectal malformations, infantile hamartoma, tail remnants and dermoid cysts, peri-rectal abscess, granuloma, osteomyelitis of sacrum, primary and secondary neurulation, caudal neural tube defect, neuroenteric cyst, neuroblastoma, neurofibroma and sub-cutaneous lipoma (Shrestha et al. 2016; Agarwal et al. 2011).

## Case presentation

A 3.5 month-old male baby presented by his parents to the pediatric surgery ward with a history of sacrococcygeal mass since birth. On clinical examination, he had a good general appearance with a soft abdomen, clear chest, open fontanels, fully conscious and usual diet with breastfeeding. The patient did not have a problem in feeding, defecation, and urination. Sacrococcygeal region revealed a mass with no discharge. Computed tomography CT scan revealed a 10 × 5 × 5 cm complex mass lesion with solid and cystic components as well as specks of calcification arising from the sacrococcygeal region extending intra-abdominally to the level of L2 vertebral body suggestive of SCT type-III (Fig. 1a ,b). The mass was excised by the impression of SCT, and sent for histopathologic examination to our department. Gross examination showed gray-white irregular tissue fragments with 5 cm in greatest fragment. The cut surface showed yellowish white homogenous and firm appearance with small cystic formation 1.0 cm in diameter (Fig. 2a). Histological examination revealed solid fragments composed of mature neural tissue comprising glial cells and astrocytes with focal hemorrhage. No evidence of other germ cell layer was found. Further sections of the specimen were grossed and submitted in additional blocks due to physician impression for teratoma and to see if there are any other tissue components. After seeing maximum slides of the tissue no other tissue components are seen. (Fig. 2b-d).

## Discussion

Congenital malformation (CM) is defined as "a permanent change produced by an intrinsic abnormality of development in a body structure during prenatal life" (Singh et al. 2014). Worldwide the prevalence of congenital malformation range from 3 to 7%. The prevalence of CM can vary with several factors contributing to its manifestation including area of study, nature of sample, ethnicity, geographical distribution and socio-economic status (Shabbir Hussain et al. 2014).

Heterotopia defined by Willis, is a general term covering all kinds of ectopic and misplaced tissues whatever their mode of genesis (Willis 1968). Heterotopic brain tissue is defined as displaced neuroglial tissue that has no connection with the central nervous system. Heterotopic brain tissue most commonly occurs in the nasal region and it is often referred to as a nasal glioma (Karma et al. 1977). The most widely accepted theory involves embryologic herniation of brain tissue through a defect in the skull, which subsequently closes and cuts off the connection between the brain and the ectopic focus (Coumou et al. 2014). Ectopic brain tissue is a rare developmental abnormality that usually has no effect on neurological development and is not associated with other congenital deformities or anomalies. The prognosis of ectopic brain tissue is good, meaning it lacks the ability to invade neighboring tissue or metastasize (Vemuganti and Shekar 1999).

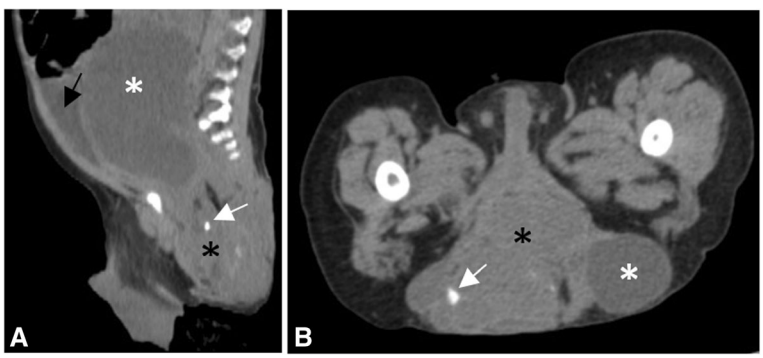


**Fig. 1** Sagittal (**a**) and axial (**b**) unenhanced CT scan through the lower abdomen and pelvic region demonstrating a (10 × 8 × 4 cm) complex mass lesion with solid (black asterisk) and cystic (white asterisk) components as well as specks of calcification (white arrow) arising from the sacro-coccygeal region extending intra-abdominally to the level of L2 vertebral body. This mass lesion is resulting in compression of the urinary bladder (black arrow) toward the anterior abdominal wall. The imaging features are suggestive of sacro-coccygeal teratoma type III

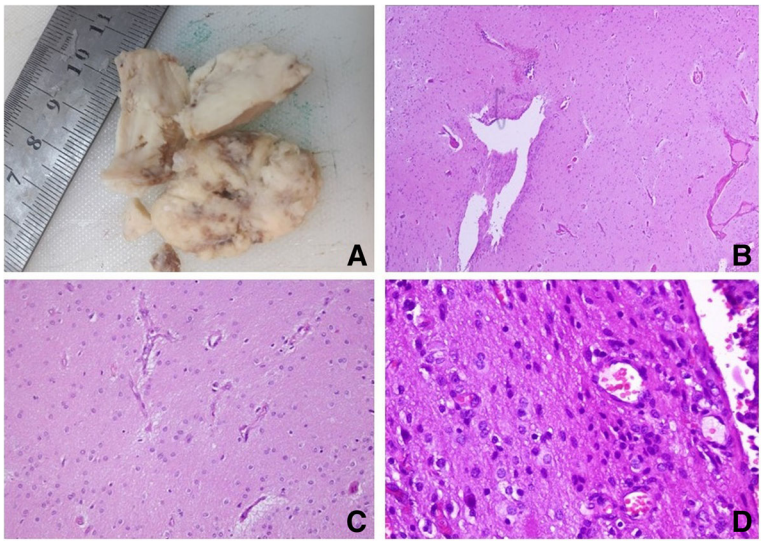


**Fig. 2 a** Gross examination showed a gray-white irregular tissue fragment**.** The cut surface showed gray to yellowish white homogenous appearance (the specimen image during additional tissue submission in the second time grossing). **b**, (4x magnification) and **c** (20x magnification), & **d** (40x magnification), Microscopic view showed solid tissue fragments composed of mature neural tissue comprising glial cells and astrocytes with no other germ cell layer component

In our case, a male neonate is presented with sacrococcygeal area mass which clinically and radiologically diagnosed as sacrococcygeal teratoma. Histopathological examination reveals only mature brain tissue which is a rare anomaly of heterotopic brain tissue in this area. To the best of our knowledge this is the third reported case of heterotopic brain tissue in the sacrococcygeal area. Two other cases of brain heterotopic brain tissue in sacrococcygeal area have been reported in female babies (Shrestha et al. 2016; Sugathadasa et al. 2013). The first reported case was a full term baby girl with sacrococcygeal area mass (Shrestha et al. 2016). The second case has been reported in 2016 in Nepal (Sugathadasa et al. 2013). In radiological examination both the cases had reported solid and cystic composition of the mass. Correspondingly, the current case radiological examination also showed mass in solid and cystic composition. The first case on CT scan report also showed few areas of calcification same findings are showed in our case by radiological examination. In sacrococcygeal region other than brain tissue one case of heterotopic adrenal gland tissue is also reported (Mandal et al. 2015). Heterotopic brain tissue other than sacrococcygeal area in other parts of body are also reported. To our best knowledge, to date ten cases of brain ectopic tissue on scalp area is reported (McGarr et al. 2001). Six cases of heterotopic brain tissue in neck and face areas are also reported (Kurban et al. 2013). In two of these reported cases the mass was confined to face with solid nature (Kern and Macdonald 1961; Kurzer et al. 1982). One case in neck area with a cystic nature which caused respiratory distress in a neonate (Robbins 3rd et al. 1985). Three other reported case were of both head and neck regions (Hendrickson et al. 1990; Lim and Capinpin 1991; Tubbs et al. 2003). Two of these cases were in compound nature of solid/cystic similar to our cases (Lim and Capinpin 1991; Tubbs et al. 2003). Other parts of body in which heterotopic brain tissue is reported such as lung (Fuller and Gibbs 1989), ectopic neural tissue in the nose (Altissimi et al. 2009), pterygopalatine fossa (Kallman et al. 1997), a case in lip (Pasyk et al. 1988) and two cases of brain heterotopic tissue in retroperitoneal region of an aborted fetus of 15 weeks gestation and another from a 3 years old boy (Hori et al. 1998). McGregor DH et al. reported three cases of heterotopic brain tissue. One case was of a child with recurrent meningitis and otitis media and two cases of elder aged people (36y and 65y) who were suffering of chronic inflammation of ear (McGregor et al. 1994). The most common challenge faced during the diagnosis of heterotopic brain tissue in any area is that mature teratoma sometimes can have predominant brain tissue. Therefore, such cases need to be carefully examined and maximum tissue should be submitted to see if there is the any possibility of other germ cell tissue. In the present case we have submitted tissue from different parts of the tissue but no other germ cell tissue was seen except the brain tissue.

Taking into consideration the mentioned reported cases of heterotopic brain tissue in the sacrococcygeal area, the diagnosis of heterotopic brain tissue in this area

should be considered in all sacrococcygeal tumors and in tumor of other parts of the body. Although it is a rare case, the diagnosis is made difficult by non-specific findings on radiological examinations and negative tumor markers.

## Conclusion

Sacrococcygeal tumor with complete brain tissue is a rare case of neonatal congenital malformations, which can be difficult to diagnose clinically and by radiologic examinations and should keep in the differential diagnosis of all sacrococcygeal tumors.

#### Abbreviations

CT: Computed Tomography; FMIC: French Medical Institute for Mothers and Children; SCT: Sacrococcygeal teratoma; UAE: United Arab Emirates; USA: United States of America

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#### Authors' contributions

JAG conceived the idea. RS was the major contributor to the writing of the manuscript. JAG and ANH diagnosed the case. JAG, RS and SR were major contributors for critically revising the manuscript for important intellectual content. HH provided the CT scan report. HG had the surgical excision of the tumor. JAG has given expert opinion and final approval of the version to be published. All authors read and approved the final manuscript.

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#### Availability of data and materials

All data generated or analyzed during this study are included in this published article.

#### Ethics approval and consent to participate

The project approved by ethical review committee of the FMIC (44-FMIC-ER-17).

#### Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report.

#### Competing interests

The authors declare that they have no competing interests.

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## Publisher's Note