



## Ovarian Tumor in A Case of Hypodontia: A Case Report

### KEYWORDS

Ovarian dermoid cyst, mature teratoma, hypodontia.

**\*Dr. Sudhir Navale**

Assistant Professor. Bharati Hospital and Medical College, Pune.  
\* Corresponding Author

**Dr. Pravin Shinde**

Assistant Professor. Bharati Hospital and Medical College, Pune.

**Dr. Priyanka Saokar**

Senior Lecturer, Dept. of Orthodontics, YCMMRDF Dental College and Hospital, Vadgaon Gupta, Ahmednagar, Maharashtra, India

**ABSTRACT** Missing teeth (hypodontia) is known to have association with ovarian tumors, particularly epithelial cell ovarian carcinoma. In the reported case, hypodontia was associated with bilateral complex cystic ovarian dermoids. Ovarian dermoid cyst or mature teratoma is the most common benign ovarian tumor in females less than 45 years of age<sup>1</sup>. Because of heterogeneous morphology and solid-cystic nature, it is sometimes difficult to differentiate them from malignant lesions. They are almost always benign and rarely show malignant transformation. We report a case of large bilateral ovarian dermoid cysts on USG & MRI in a patient who underwent ovarian screening after dental anomalies were found.

### INTRODUCTION

Hypodontia, defined as congenital absence of one to five teeth, is a dental disability that affects a patient's function and esthetics. Mutations of MSX1 and PAX9 genes are most commonly associated with hypodontia and same genes have role in tumor growth inhibition.

Ovarian dermoid cysts tend to be slow-growing tumors and are often asymptomatic. Cystic teratomas make up approximately 15 to 25 percent of ovarian neoplasms. They are composed of well-differentiated derivatives of 3 germ layers, ectoderm, mesoderm and endoderm. Cystic teratomas are more commonly seen in active reproductive years but can occur at any age and are frequently seen in postmenopausal women<sup>1</sup>.

### CASE REPORT

A 25 year old asymptomatic female underwent ultrasound screening of the abdomen & pelvis to look for congenital abdominal organ anomalies; after dental anomalies were found in orthodontic clinic. She had congenitally missing mandibular right lateral incisor along with sella turcica bridging. Fig.1 and Fig.2.

Ultrasonography revealed a multiloculated, complex left ovarian cyst of 17.4 x 10.4 x 18.7 cm with mural solid areas in one locule (Fig.3). A possibility of cystadenoma or cystadenocarcinoma was raised, because of large cystic component with solid mural nodules.

Fig. 1 and Fig. 2



Fig. 3



Her CA-125 was 15.12 (normal < 35 U/ml). Pelvic MRI was performed for further characterization of the lesion.

MRI showed a predominantly cystic lesion (78 x 143 x 186 mm in AP, transverse and cranio-caudal dimensions) in the pelvis extending into the lower abdomen. It appeared hypointense on T1W images and hyperintense on T2W & STIR images (Fig. 4, 5 & 6). Multiple thin septae were seen within the lesion. A focal hyperintense area was seen within it on T1W images appearing hypointense on fat sat images representing fatty tissue. On post-contrast images the septae showed mild enhancement. Left ovary could not be seen separately from the lesion.

Another lesion (33 x 47 mm) was seen within the right ovary with fatty content. A focal solid hyperintense area was seen in addition to the fatty tissue along its inferior aspect on T1W images, appearing hyperintense on T2W images. On post-contrast study, this solid component showed heterogeneous enhancement (Fig. 7, 8 & 9).

MR findings were consistent with bilateral ovarian dermoids.

Fig. 4



Fig. 5

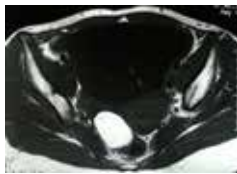


Fig. 6



Fig. 7

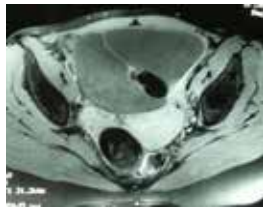


Fig. 8

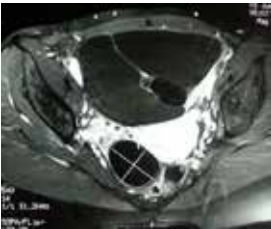


Fig. 9



The lesions were excised surgically and sent for histopathology. Histopathological report was consistent with bilateral ovarian benign cystic teratomas.

## DISCUSSION

Tooth morphogenesis is a complex process that involves epithelial-ectomesenchymal interactions. Numerous transcription factors, growth factors, and their receptors, as well as extracellular matrix components, have been associated with early tooth development.<sup>2,3</sup> The genetic basis of tooth development is supported by the identification of mutations in genes that participate in dental development (MSX1, PAX9, AXIN2)<sup>4,5,6</sup>. More recently, a mutation in the AXIN2 gene was identified in families with oligodontia and colorectal cancer, suggesting that tooth agenesis might be an indicator of colorectal cancer susceptibility.<sup>7</sup> Hypodontia may also be present in certain syndromes (Down syndrome), 26 ectodermal dysplasia.<sup>8</sup> New researches also reported a higher presence of cancer in family members of patients with hypodontia.<sup>9</sup>

The terms dermoid and teratoma are not synonymous. A dermoid tumour contains only ectodermal elements, whereas a teratoma can contain any or all of the three germ cell layers: ectoderm, endoderm, and mesoderm. Benign cystic dermoid tumours constitute about 98% of all teratomas. The rest are immature and specialized varieties, or rare solid mature teratomas in which tissues from all three germ cell lineages form a heterogeneous compilation. Mature teratoma, typically cystic tumours containing varying amounts of hair, sebum, and tooth elements, occur in young women during their reproductive years. The lesions are bilateral in 10% to 25% of cases.<sup>10</sup>

Cystic teratomas demonstrate a broad range of findings on imaging due to varying degree of tumour content. Several diagnostic imaging modalities, such as transabdominal and endovaginal ultrasound with Colour Doppler assessment, CT and Magnetic resonance imaging, can be used to help discover, characterize, and differentiate ovarian masses.<sup>11</sup> On ultrasound, they may contain an echogenic tubercle known as Rokitansky nodule. If bone and teeth are present, they tend to be located in this nodule. Hair is seen as echogenic bands. Acoustic shadowing may obscure the posterior wall of a large mass -- "tip of the iceberg" sign.<sup>6,12</sup>

MRI is a useful adjunct in assessment of ultrasound indeterminate lesions. On MR, the sebaceous component will have high signal on T1 and T2W sequences, but T2 signal can be more variable in signal intensity. Fat suppression techniques will help differentiate fat from hemorrhage with lesions that are high on T1W sequences.<sup>13,14</sup> Teeth or bone will be low signal on all sequences. Treatment involves laparoscopic or surgical resection.

Complications of mature cystic teratomas include torsion, rupture, infection and malignant transformation. Malignant transformation is uncommon and is seen in approximately 2 percent of cases, usually in older women.<sup>1</sup>

More than 300 genes are involved in odontogenesis, and mutations in several of these genes have been linked with hypodontia.<sup>15,16</sup> The genes that control the development of teeth also have important functions in other organs and body systems. Therefore, it is plausible to assume that a genetic mutation resulting in hypodontia also may cause abnormalities in other parts of the body. Congenital absence of permanent teeth has direct clinical (visual) implication, and early evaluation of hypodontia phenotype could help us to identify patients at risk for epithelial cell ovarian cancer and serve as a possible marker for abnormalities in other systems. Early screening regimens in cases of hypodontia should result in identification of these high risk individuals leading to early diagnosis.

## REFERENCE

- Shia Salem, The Uterus and Adnexa In Diagnostic Ultrasound by Carol M Rumack, Stephanie R 1997, 2nd edition; Volume 1: 557-558. | 2. Thesleff I, Nieminen P. Tooth morphogenesis and cell differentiation. *Curr Opin Cell Biol.* 1996;8:844-850. | 3. Miletic I, Sharpe PT. Normal and abnormal dental development. *Hum Mol Genet.* 2003;12:69-73. | 4. Vastardis H, Karimlux N, Guthua SW, Seidman JG, Seidman CE. A human MSX1 homeodomain missense mutation causes selective tooth agenesis. *Nat Genet.* 1996; | 5. Stockton DW, Das P, Goldenberg M, D'Souza RN, Patel PI. Mutation of PAX9 is associated with oligodontia. *Nat Genet.* 2000;24:18-19. | 6. Mostowska A, Biedziak B, Jagodzinski PP. Axis inhibition protein 2 (AXIN2) polymorphisms may be a risk factor for selective tooth agenesis. *J Hum Genet.* 2006;51:262-266. | 7. Lammi L, Arte S, Somer M, et al. Mutation in AXIN2 cause familial tooth agenesis and predispose to colorectal cancer. *Am J Hum Genet.* 2004;24:1043-1050. | 8. Kere J, Srivastava AK, Montonen O, et al. X-linked anhidrotic (hypohidrotic) ectodermal dysplasia is caused by mutation in a novel transmembrane protein. *Nat Genet.* 1996;13:409-416. | 9. Kucher EC, Lips A, Tannure PN, et al. Tooth agenesis association with self-reported family history of cancer. *J Dent Res.* 2013;92:149-155. | 10. SeungEun Jung, Jae Mun Lee, Sung EunRha et al. CT and MR Imaging of Ovarian Tumors with Emphasis on Differential Diagnosis. *Radiographics* 2002; 22: 1306-25. | 11. Radiology rounds, Michael K McLennan, Myles Margolis. *Canadian Family Physician* 1994 October; 40: 1-4. | 12. Sutton CL, McKinney CD, Jones JE, Gay SB. Ovarian masses revisited: radio-logic and pathologic correlation. *Radiographics* 1992;12:853-77. | 13. Sung EunRha, Jae Young Byun et al. Atypical CT and MRI Manifestations of Mature Ovarian Cystic Teratomas. *September 2004, Volume 183, Number 3.* | 14. Elkhaili Alaylahi, MD, Rachida Dafiri. *Applied Radiology: Bilateral Ovarian Dermoid Cyst.* October 12, 2005. | 15. Thesleff I. The genetic basis of tooth development and dental defects. *Am J Med Genet A* 2006;140(23):2530-5. | 16. Tooth and Craniofacial Development Group of the Developmental Biology Programme, Institute of Biotechnology, University of Helsinki. Gene expression in tooth development. Available at: "http://bite-it.helsinki.fi". Accessed Jan. 5, 2007. |