Dermoid Cyst and its bizarre presentation

Pramila Pradhan,¹ Meeta Thapa¹

¹Department of Obstetrics and Gynaecology, Nepal Medical College Teaching Hospital, Jorpati, Kathmandu, Nepal.

ABSTRACT

Dermoid cysts or mature cystic teratoma are the most common type of ovarian germ cell tumor usually occurring in the reproductive age group. They are commonly unilateral and 10-12 % can be bilateral. They are commonly multicystic and contain sebaceous fluid as well as tissue of three germ cell layers e.g. ectoderm (skin, hair, brain), mesoderm (muscle, fat, teeth, bone, and cartilage) and endoderm (mucious and ciliated epithelium). These teratomas usually arise from the gonads but has been found anywhere in the body. Their site, size and clinical presentations are extremely variable causing confusion with medical and surgical diseases, acute emergency due to rupture and torsion, bowel and bladder injuries, and pregnancy etc. The malignant teratoma occurs at 3-4% only among ovarian carcinoma. The malignant transformation in benign cysts is a rare occurrence with 1-2% cases and squamous cell carcinoma being the commonest. This review is undertaken to study the different presentations produced by these tumors.

Keywords: bizarre; cyst; dermoid.

INTRODUCTION

Mature cystic teratomas commonly called dermoid cysts, are the most common benign germ cell tumors of ovary, comprising up to 30% of all ovarian tumours.¹ They are encountered predominantly in women in their second and third decades of life but can occur in any age group 1-96 years.² Histologically, they display a varying admixture of elements of one or more of the three germ cell layers: ectoderm (skin, brain) mesoderm (muscle, fat, teeth, bone and cartilage) and endodermal (mucinous or ciliated epithelium gastrointestinal, bronchial, thyroid tissue).³ Pathologically, teratomas are classified into three groups: Mature (cystic/solid, benign), Immature (malignant) and Monodermal (highly specialized e.g. Struma ovarii, carcinoid tumors, neural tumors). Teratomas most commonly arise in the gonads but have also found in the anterior mediastinum, retro-peritoneum and gastrointestinal tract.⁴ Despite the benign nature of these neoplasms, considerable interest have been paid on them because of unusual histogenesis caused by the totipotency of these tumors that may give to any body structure or tissues and even the structure similar to fetus. ⁵ Despite a number of conflict theories to explain the histogenesis of teratomas, the most probable ones are misplaced blastomere and pathenogenetic development of a germ cell. Whatever the reason for the development of teratomas may be, these tumors seemingly arise from the totipotential embryonic cells broken away from the normal developmental pathway to form the various fetal tissues.⁶ The majority of teratomas may be symptoms free but giant ovarian teratomas commonly present with acute abdominal pain caused by adnexal torsion, rupture or infection or abdominal distention due to the rapid growth of a large, unilateral tumor undergoing capsular distention, hemorrhage or necrosis.⁷ The mature teratomas sometimes may be adherent to small and large bowel causing intestinal obstruction or fistula
Malignant transformation of the mature elements within the dermoid cyst is a rare complication, occurring in only 1-2% of cases with squamous cell carcinoma being the most common type (80-90%) followed by adenocarcinoma, Carcinoid, Sarcoma, Osteosarcoma. At ultrasound mature teratomas are characterised by echogenic sebaceous material and calcification. At Computed Tomography (CT) fat attenuation within a cyst is diagnostic. CT and Magnetic Resonance Imaging (MRI) are straightforward as these modalities are more sensitive for fat within the cyst which is diagnostic for mature teratoma. Their clinical features vary from completely asymptomatic to bizarre features confusing with medical, surgical diseases, acute emergency due to rupture or torsion of the cyst, bowel or bladder injuries and malignant transformation.

This article tries to present different presentations, some of which are bizarre nature encountered with mature teratomas.

**Size**

The size of a dermoid cyst may be extremely variable. They can be found accidentally when they measure just around 1cm and are situated inside the ovary without causing ovarian distortion. (Figure. 1).

They have a growth rate of 1.8mm/year in premenopausal women and rarely grow larger than a melon size, but they can also have gigantic dimensions of 30-40cms. (Figure 2). The tumor tend to enlarge until specific organs are functionally influenced or incidentally discovered by ultrasonography. Massive ovarian tumors can mimic actites and usually present with obvious abdominal distention but in the presence of severe obesity they may be overlooked. There have been reports in both the medical and lay press of surgical extirpation of simple fluid filled cysts weighing as much as 300lbs and draining 80 litres of fluid. An ovarian teratoma of 303.2 pounds (134.7kg) removed enbloc has been described in a bed ridden lady for the previous 2 years and house bound for 6 years due to enlarging ovarian dermoid cyst. Despite the large size, there is a one third chance of finding a malignant suggesting that all of these large masses should be removed intact whenever possible.

**Site**

Dermoid cysts occur most commonly in the ovary but the cases of extragonadal origin have been described e.g. anterior mediastinum, retroperitoneal, sacrococcygeal area, head, neck and central nervous system (CNS). Dermoid cysts have been reported throughout the gastrointestinal (GI) tract and associated organs include caecum, rectum, pancreas. In the GI tract mesenteric dermoid cysts are the most common site for ovarian teratoma. Omentum is also the most common extragonadal site of dermoid.
cyst. The omentum, because of its special role in intra-abdominal inflammatory defence process, is probably the main location for secondary implantation for the tumor.21,22

Parasitic dermoid cyst

Parasitic dermoid cyst is extremely rare entities and their actual incidence is reported to be 0.4% of all ovarian teratoma.23 Several theories exist to explain their occurrence. The first theory proposes the development within a supernumerary or ectopic ovary which may occur due to implantation of ovarian tissue after surgical procedure or inflammation such as pelvic inflammatory disease. Supernumerary ovaries may also occur as a result of abnormal arrest of germinal cells into the dorsal mesentery during their embryonic migration.24 Another theoretical mechanism is auto amputation and re-implantation of the dermoid cyst as a result of torsion. If the torsion of the tumor is subacute, an inflammatory response may occur which causes the tumor to become adherent to the surrounding structures and neovascularization may occur. The tumor may become parasitic by detaching itself from its original blood supply.25 Pouch of Douglas seems to be the best location for the parasitic dermoid.22 Recently a new site for parasitic dermoid cyst on the reflection of utero-vesical pouch onto anterior abdominal wall which is known anatomically as the median umbilical fold have been reported.25,26 Some of these parasitic dermoid cysts have been found to incidentally contain mobile spherules. The reason for the formation of such large spherules is still unknown, although it is proposed that hypovascularity could be a possibility.27,28 The clinical presentation of the reported parasitic teratomas of the pouch of Douglas tends to mimic those of omental teratomas, namely abdominal pain. The radiation of pain of the patients back is likely from lumbosacral neuropathy, due to the compression of the lumbosacral plexus.27

Torsion

Torsion of the pedicle is reported to be the most frequent complication of ovarian teratoma, occurring in 16.1% of cases.22 Torsion interferes with the blood supply of the involved organs. Venous congestion and aseptic inflammation of the tumor wall may thus result. In acute torsion, the tumor undergoes necrosis and subsequently atrophies due to ischemia. In subacute or chronic torsion the tumor may become adherent to adjacent structures with new collateral circulation formed. Infrequently, the tumor completely detaches from its pedicle, thus resulting in a parasitic dermoid cyst. Dermoid cyst is an infrequent cause of abdominal and flank pain, when it does occur, it is due to mass effect or ovarian torsion. The pain is related to the ischemic complication of the enlarged ovary twisting around its pedicle.29

Chemical and Granulomatous peritonitis

Inadvertent dermoid cyst rupture may result in chemical peritonitis (3-7%), and surgical treatment may become necessary.28,30 Rupture of the cyst with spillage of its fluid content may be deleterious with teratomas. Chemical peritonitis and granuloma formation with intestinal obstruction have been reported after laparoscopic removal of benign cystic teratomas due to spillage.29 Adequate intra-abdominal isolation within an endobag to avoid the most common event of spillage during laparoscopic removal of dermoid cyst is now advocated by several investigators.26,31

A retrospective study covering 20 years and covering 26 cases of intra-peritoneal spillage in 314 patients identified 2 patients with post-operative granulomatous peritonitis, giving an incidence of chemical peritonitis 8%.22 However the data from another retrospective study of 324 patients who underwent laparoscopic cystectomy and suffered spillage, with irrigation of the abdominal cavity documents a lower incidence (0.3%).33 However, prospective controlled clinical trials with a large number of patients are necessary to compare conventional methods with laparoscopy in cases with teratomas and to assess more rare events like undetected malignancy, clinical peritonitis or excess adhesion formation.

Small bowel obstruction with ovarian teratoma

Ovarian teratomas are usually asymptomatic until they reach a significant dimension. Pressure effect, torsion and rupture of an ovarian cyst may present as an acute abdomen. A case has been reported in an elderly lady with small bowel obstruction due to a very large ovarian teratoma adherent to the intestine causing intestinal obstruction.34 Acute intestinal obstruction is a very rare complication of ovarian teratomas. It will mainly occur when a loop of a small bowel becomes adherent to the cyst and twist with the torsion of the cyst requiring intestinal resection.35,36

Fistula formation with bowel

Ovarian teratoma fistulating to the sigmoid colon causing great abdominal distention is an extremely rare case. Although there are numerous differential diagnoses, most of the diagnoses would become apparent after radiological work-up. Fistulation is a rare complication of mature cystic teratoma and urinary bladder is the most commonly affected organ.37,38 Malignant transformation is not a pre-requisite for fistulation. Inflammation related to previous sub clinical leakage
of the cyst content is believed to be the etiology of fistulation in some of the previous reports. Both inflammation and direct infiltration by malignant cells had contributed to the formation of fistulation in a case report of a giant gas filled abdominal mass in an elderly female. The presence of significant tumor bulk around the area of fistulation suggest tumor invasion being the pre-dominant factor. Malignant transformation arising from ovarian teratoma can rarely present as a surgical emergency with gross abdominal distension, resembling bowel pathology such as giant colonic diverticulum and duplication cyst. A case of benign cystic teratoma with fistula into the small and large bowel has been reported. A 10cm dermoid cyst perforating into the small and large bowel is described in a 25 year old lady. A primary large bowel repair, small bowel resection with enblock left salphingo-oophorectomy and small bowel side to side re-anastomosis saved the lady. Awareness to the clinical and radiological findings in these cases is important to avoid further complications.

Malignant transformation in a dermoid cyst

Dermoid cysts contain tissue from all three germ cells with teeth, bone and neural tissue being found. Secondary malignancies may develop in any of the tissue derived from the three germ layers. Malignant transformation of the mature elements within the dermoid cyst is a rare complication, occurring in only 1-2% of cases with squamous cell carcinoma being the most common type (upto 80-90%) followed by adenocarcinomas, carcinoid, neuroblastomas, sarcoma, dermoid cyst. A combination of different malignancies arising in the same dermoid cyst or the incidence of more than one malignancy arising from one ovary is exceptionally rare. But different combinations have been reported in the literature. Hanada et al described the multiple malignancy arising from a dermoid cyst of the ovary namely squamous cell carcinoma and myxoid fibrous histiocytoma. Arora and Haldane reported a case of carcino-sarcoma arising in the dermoid cyst of the ovary comprising of adenocarcinoma and leiomyosarcoma. A unique combination of squamous cell carcinoma and osteosarcoma arising in mature dermoid cyst of the ovary have been reported. The pathogenesis of malignant transformation of one somatic cell type to another is not clearly understood. It seems that mature dermoid cyst most likely to undergo transformation of totipotential germ cells towards the formation of squamous cell carcinoma that might have later dedifferentiated into osteosarcoma. A large mature cystic teratoma containing carcinomatous and sarcomatous elements has been reported. Sarcoma alone or in combination with squamous carcinoma has been described arising in a mature cystic teratoma but a case of sarcoma arising in associated with adenocarcinoma is very rare. Adenocarcinomas are the second most common malignancies arising within the dermoid cyst.

It is suggested that age is not necessary a prognostic indicator but may be useful in terms of raising suspicion of malignant transformation when an older women presents with what appears to be a benign dermoid cyst. Other risk factors include tumor diameter greater than 10cms and rapid growth. Al Rayyan et al in their publication concluded that regardless of the type of malignancy, the tumor size, the extent of the disease and the patient’s age were the major factors governing the survival in these patients. It is recommended that a higher index of suspicion of malignancy is needed in managing dermoid cysts occurring in patients over the age of 45 years especially if rapidly growing or very large tumors greater than 10 cm. Kelly and Scully reported that thickening of the cyst wall and adherence to surrounding structures should raise high index of suspicion for malignancy in a dermoid cyst.

Stromal tumors arising from the mesodermal germ cells of a mature dermoid cyst are rarer than their epithelial counterparts but have been reported. Ngwalle et al described the typical case occurring in a post-menopausal women with an osteosarcoma arising in benign dermoid cyst. A combination of different malignancies arising in the same dermoid cyst or the incidence of more than one malignancy arising from one ovary is exceptionally rare. But different combinations have been reported in the literature. Hanada et al described the multiple malignancy arising from a dermoid cyst of the ovary namely squamous cell carcinoma and myxoid fibrous histiocytoma. Arora and Haldane reported a case of carcino-sarcoma arising in the dermoid cyst of the ovary comprising of adenocarcinoma and leiomyosarcoma. A unique combination of squamous cell carcinoma and osteosarcoma arising in mature dermoid cyst of the ovary have been reported.
and may display gastrointestinal phenotypes but lack the malignant stroma.  

The prognosis of patient with malignant transformation in teratoma is very poor with women dying within a year. Occasionally cases with prolonged survival have been reported. Poor prognostic factors include tumor dissemination, cyst wall invasion, ascites, spontaneous or accidental rupture, adhesions and tumor type other than squamous carcinoma.

It is histologically difficult to diagnose this disease pre-operatively as neither serum tumor markers nor imaging with ultrasound or CT reliably predicts the specific diagnosis. CA19-9 is a tumor associated glycolipid, frequently elevated in the serum of patients with gastrointestinal malignancies particularly of the pancreas. It is present in relatively high concentration in prostatic, gastric, amniotic fluid and excretions of pancreas and duodenum in individuals. CA19-9 could be increase in upto 50% of cases of dermoid cysts with malignant changes. Modern imaging techniques such as MRI shows the presence of solid or necrotic contrast-enhancing areas that may extend transmurally and show evidence of adherence to surrounding structures. Mature cystic teratoma of ovary frequently and show evidence of adherence to surrounding areas that may extend transmurally such as MRI shows the presence of solid or necrotic contrast-enhancing areas that may extend transmurally and show evidence of adherence to surrounding structures. Mature cystic teratoma of ovary frequently contain intestinal type epithelium, but they are rarely associated with complete intestinal wall. The association of mature cystic teratoma with mucinous cystadenoma is not unusual. However, the pathogenetic relationship between these two lesions remain unanswered. A mature cystic teratoma containing a complete colonic wall in continuity with an endocervical type mucinous cystadenoma is reported. Both the mucinous cystadenoma and the colonic wall showed the typical histopathological and immuno-histochemical patterns of classical mucinous cystadenoma and normal colonic wall. The microscopic and immuno-histochemical patterns of the transitional zone between colonic wall-like structure and mucinous cystadenoma stained positive for chromogranin suggesting that the mucinous cystadenoma most likely originated from colonic epithelium of the mature cystic teratoma.

Infection

Mature cystic teratoma may be complicated by torsion, rupture, chemical peritonitis and malignant change but is rarely complicated by infection. Infection occurs in approximately 1% of mature cystic teratoma, the infecting organism most likely being coliforms. Actinomycosis, Brucella, Schistosomiasis, Salmonella has also been reported. It is more commonly seen in reproductive age but seen in post-menopausal women also. A tubo-ovarian abscess following a dilatation and curettage (D&C) procedure in the setting of an ovarian dermoid cyst have been reported.

Homunculus in dermoid cyst

In Latin, Homunculus means a structure resembling a miniature human body to designate a human being not produced by pregnancy and this definition is used in morphological entities. Such fetus like structure (homunculus) can be found in mature cystic teratoma. In some cases the cephalic region is more developed with cranium and cerebral cortical tissue. In some cases the caudal region including the lower extremities is typically more developed than the cephalic portion. In a case reported, the radiographic examination of the homunculus revealed a calcific density in the cephalic portion resembling temporal bone, zygomatic arch, temporomandibular joint and mandible. The caudal portion showed a tubular bony structure. This tumor should be distinguished from the more highly developed fetus-in-situ, a malformed parasitic monozygotic twin that is found inside the body of a living child or adult.

Multiple recurrence of dermoid cyst

In the literature it is observed that mature cystic teratoma can show recurrence with a rate of 3-4%. In different reports it is emphasized the especially cysts with diameter less than 2cm can be ignored during preoperative transvaginal ultrasonographic imaging but these cysts can be later be diagnosed as recurrence. It is realized that the recurrence rate was higher in the patients who were young at first diagnosis. At the same time, bilaterality and multi-locularity were observed more often in the recurrent cases. The recurrence occurs nearly 1-15 years after the first operation and the recurrence rate was 2-5% after ovary protected surgery. The regular follow-up of young patients has a clinical importance in the early detection of the recurrent masses and during the surgeries of the recurrent cases, the surgeon should be careful about the bilaterality or multilocularity of the tumor.

Virilizing ovarian dermoid cyst with leydig cells

Ovarian tumors causing virilization are exceedingly rare and mostly occur in post-menopausal women. In fact, there are no reported cases of virilization from a testosterone producing ovarian dermoid in the adolescent female age group. A case has been reported in 12 year old girl who has a large 35 X 19 X 12 cm ovarian mature cystic teratoma arising from her right ovary. The patient also displayed evidence of masculinization demonstrated by a deepening voice and citoromgaly. The dermoid was producing large amounts of testosterone from a nest of leydig cells found pathologically in the mass.
teratomas can produce active hormones, albeit rarely. This is a finding important to consider when ovarian cystectomy is performed for removal of a benign cystic teratoma.

CONCLUSION

The cornerstone of the study of ovarian teratoma is that the evaluation of possible malignancy, accidental rupture/ hemorrhage, rapid growth, doubtful infiltration, large size, origin and adjacent structures is based on detailed preparation. Despite the large size, there is one third chance of finding a malignancy, suggesting that all of these large masses should be removed intact wherever possible. Prospective controlled clinical trials with a large number of patients are necessary to compare conventional methods with laparoscopy in cases with teratomas and to assess more rare events like undetected malignancy, chemical peritonitis, or excess adhesion formation.

REFERENCES


