

## Giant mucinous cystadenoma in a young patient. A case report

S. GORGONE, C. MINNITI, A. ILACQUA, M. BARBUSCIA

**SUMMARY:** Giant mucinous cystadenoma in a young patient. A case report.

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*The Authors describe the clinical case of a seventeen-year-old girl who presented with abdominalgia, fever, nausea and vomiting. During surgery it proved necessary to remove not only the appendix but also a voluminous mucinous cystadenoma of the ovary. The authors take this observation as a starting point for a description of the pathophysiological and clinical findings of these neoplasms.*

**RIASSUNTO:** Cistoadenoma mucinoso gigante in una giovane paziente. Su un caso clinico.

S. GORGONE, C. MINNITI, A. ILACQUA, M. BARBUSCIA

*Gli Autori presentano il caso clinico di una diciassettenne con una sintomatologia caratterizzata da dolore addominale di tipo crampiforme, iperpiressia, nausea e vomito alimentare. Durante l'intervento chirurgico si dovette procedere, oltre che all'appendicectomia, all'asportazione di un voluminoso cistoadenoma mucinoso dell'ovaio.*

*Gli Autori prendono lo spunto da questa osservazione per descrivere le caratteristiche fisiopatologiche e cliniche di tali neoformazioni.*

KEY WORDS: Cystadenoma - Ovarian cancer.  
Cistoadenoma - Tumore ovarico.

### Introduction

Mucinous cystadenomas are tumours deriving from the surface (coelomic) epithelium of the ovary due to abnormal lack of regression of Müllerian cells. According to some authors, they account for 25% of all ovarian neoplasms; mucinous cystadenomas are also the most frequent form of cystic neoplasm to affect this organ.

While tending to occur between the fourth and the fifth decades of life, they can arise at any age, even though they are particularly rare during and before puberty (1,2) and in post-menopausal women.

These tumours are malignant in 15% of cases, and bilateral in 5% (3,4).

### Case report

The authors present the clinical case of a seventeen-year-old girl, B.M., referred to them directly from the Emergency Room (ER). The patient reported the onset of the symptoms about 12 hours prior to her admission when she had begun to present abdominalgia, fever, nausea and food vomiting. The symptoms failed to regress following the assumption of drugs prescribed by her family physician. For this reason, she went to the ER from where she was sent to us with a diagnosis of appendicitis.

When she arrived, her abdomen was tractable and tender only in the right iliac pit; in the same area, deep palpation elicited pain. An area of dullness was found on percussion, and borborygmi could be heard on auscultation of the abdomen. McBurney and Blumberg's signs were found to be weakly positive.

This clinical picture confirmed the diagnosis of the ER and the patient underwent surgical operation. After right pararectal incision and opening of the peritoneum, it was possible to observe the wall of a voluminous cystic neoplasm arising from the right ovary. The laparotomic incision was widened to allow complete removal of the cyst, which measured 20x14x6.5 cm, and homolateral salpingectomy (Fig. 1). The surgical exploration also revealed a hyperaemic appendix, making it necessary carry out an appendectomy.

The patient was discharged five days later in good conditions, with no fever and canalized.

Histological examination of the operative specimen revealed it to be a "multichambered cystic neoplasm". On incision, some cavities leaked a serum-like liquid, others were covered by a gelatinous, mucoid material. A whitish solid area (2x1 cm) was also ob-

University of Messina  
Chair of General Surgery  
(Head: Prof. S. Gorgone)  
Chair of Digestive Tract Surgery  
(Head: Prof. M. Barbuscia)

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**Fig. 1 - Open mucinous cystadenoma.**

served. On the basis of the histological report, a diagnosis of “multilocular mucinous cystadenoma” was made.

## Discussion

The pathogenesis of these cystic tumours is uncertain. Various theories exist (5-7), the most accredited of which suggests that they derive from inclusions and invaginations of the surface (coelomic) epithelium of the ovary that are produced during the repair of the follicle wall after ovulation. It is thought that inclusion cysts form initially and that the various subtypes of cystic neoplasm develop subsequently as a result of metaplasia of the included coelomic epithelium, or that they arise from the entodermal epithelia of teratomas; finally, an alternative theory is that they originate from remains of the Wolffian epithelium, often present in the ovary.

Macroscopically, these tumours have the appearance of cysts, are generally monolateral and almost always pedunculate. They tend to reach considerable dimensions (some even growing so large that they occupy the whole pelvic cavity and abdomen), giving rise to respiratory and circulatory problems, oedema, etc. (8, 9). There are literature reports of tumours reaching 16.4 kg. The complexion of the cyst is greyish white, if the wall is thick, and slightly yellow and translucent if the wall is thin. On incision it can prove to

be unilocular or have multiple cavities; the content is a dense, gelatinous liquid, clear but, at times, reddish or dark red if mixed with blood; if infected, the liquid can appear yellowish, like pus. This liquid is primarily made up of a PAS-positive glycoprotein (pseudo-mucin); it contains a number of enzymes (amylase, lipase, maltase, invertase, lactase, papaine) and minerals, particularly potassium. High quantities of oestrogens and 17-chetosteroids and traces of pregnandiolum have also been found.

Histologically, the coating epithelium appears cylindrical and mono- or multi-stratified, with cells presenting a clear cytoplasm and hyperchromatic nucleus at the base. Its appearance is similar to that of the epithelium that covers the endocervix or to that of the intestinal wall. Alternatively it can appear cuboid and mono- or multi-stratified; this would seem to be due both to the pressure exerted by the liquid inside the cyst, and to the variations of the functional activity of the epithelium, that has therefore an elaborative phase and a secretory phase. More recently, this diagnosis has been confirmed using immuno-histochemical techniques (CD24).

Clinically, because these tumours are slow-growing, the symptoms tend to be mild. They are often discovered during periodic gynaecological examinations or by the patient herself who, palpating her abdomen, discovers a mass that she has never felt before (10-12).

If present, the symptomatology is always aspecific: vague abdominal pain on the affected side, difficult digestion, meteorism, rare signs of bladder and/or rectal compression. Menstrual disturbances are infrequent. Occasionally these tumours can present complications (torsion, rupture, haemorrhage and infection), that sometimes constitute the first symptom of the illness. It is then that there appears a more severe symptomatology, still aspecific, in the form of abdominalgia simulating acute or sub-acute abdomen due to peritoneal irritation (13, 14).

The objective examination of the abdomen will reveal the presence of a tumour mass, of varying dimensions, soft or tense-elastic consistency, and non-constant form. On percussion, the mass will produce a dull sound that is not modified when the patient changes position. Also present is the sign of Fourestier: a clear sound in the lumbar region, due to the fact that the cyst pushes the intestine in the lumbar pit.

The above-described clinical picture can be clarified and/or confirmed through instrumental diagnostic methods, in particular by echotomography, also performed using a vaginal probe, that makes it possible, in 85-95% of the cases, to observe the presence or absence of an ovarian swelling, to establish its diameter and content (liquid, solid or mixed), as well as so-

me internal features of the tumour mass (unilocular vs multilocular, thin or thick septi, presence of inner proliferations of bony tissue, etc.).

When ultrasound investigation fails to resolve the diagnostic doubt, CT scans (preferably with the use of contrast enhancement) can be helpful, as can explorative laparoscopy (essential when the data obtained are not still enough to justify recourse to laparotomy and when the ovarian mass has a small diameter). Furthermore, through laparoscopy it is possible to sample the peritoneal fluid, to perform a lavage of the pelvis or to aspire the possible ascitic fluid for subsequent cytological examination.

The differential diagnosis of this condition is not straightforward and it must take into account, mainly, the following clinical conditions:

1. uterus increased in volume and modified due to pregnancy;
2. uterine fibromyomatosis;
3. retroverted and fixed uterus;
4. pelvic endometriosis;
5. voluminous tubaric inflammatory harvest (sactosalpinx);
6. extra-uterine pregnancy;

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7. full bladder due to urinary retention;
8. some intestinal pathologies: dolichosigmoid; ptotic caecum; faecaloma; diverticulitis; Crohn's disease; cancer of the sigma or caecum;
9. peri-appendicular abscess;
10. pelvic kidney;
11. pelvic retroperitoneal tumour;
12. obesity or meteorism;
13. ascites of non genital origin;
14. mesenteric cyst or cyst of the urachus;
15. pelvic echinococcosis.

## Conclusions

The diagnosis of ovarian cystadenoma is often difficult as different clinical conditions can disguise the clinical picture, not always univocal, of this pathology. A precise diagnosis allows a planned, elected surgical operation, without the risk of running into sometimes serious complications. Regular gynaecological examinations after the menarche, scheduled together with the specialist, could facilitate the achieving of this objective.