An ultrasonographic approach to unusual peritoneal disease (mesothelioma, plasmacytoma and acute plasma cell leukemia)

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Learning objectives

The aim of the poster is: 1) to describe the ultrasonographic findings of diseased peritoneum in the cases of mesothelioma, plasmacytoma and acute plasma cell leukemia; 2) to become familiar with the potential role of conventional ultrasound in detecting signs of primary or secondary peritoneal disease (mesothelioma vs plasma cell leukemia and plasmacytoma) during a routine / general abdominal sonogram.

The clinical and radiologic presentation of this three entities is nonspecific, among the most frequently reported symptoms being abdominal pain, abdominal swelling, anorexia and marked weight loss [1-4]. Thus, the role of the US in performing a quickly and efficiently survey of the peritoneum, peritoneal cavity and the solid organs, that would guide the clinician in the direction of a possible diagnosis of peritoneal involvement in the case of different pathologies.
Background

Although the traditionally imaging modality of choice used in assessing pathologic conditions of the peritoneum and peritoneal cavity is the CT, some signs of peritoneal disease can be encountered unexpectedly (incidentally) during abdominal ultrasound (US) performed to evaluate localized abdominal or pelvic pain or/ and discomfort, that represent the clinical presentation of a wide range of diseases [1,10,11].

Thus, the understanding of characteristic appearances will aid the radiologist in the diagnosis of this very rare and sometimes overlooked causes of nonspecific abdominal pain.

Malignant mesothelioma is a rare, fatal neoplasm arising from the serous surfaces, involving the peritoneum in 33% of the cases, either solely or in combination with pleural involvement [1, 2].

Peritoneal mesothelioma is diagnosed in advanced stages in most cases, in part because of the usually long latent period and secondly because the common presenting symptoms of weight loss, malaise, abdominal fullness and discomfort are mild and nonspecific. This disease usually remains confined to the peritoneal cavity for most of its natural history. Typical growth pattern of peritoneal mesothelioma is locally expansive masses. Although hematogenous or lymphatic metastasis is unusual, there have been reported parasternal, retroperitoneal, mediastinal, axillary, supraclavicular, and cervical lymph nodes; lung, bone, liver, and umbilical metastases [2,5,6].

Even with multitechnique therapy the prognosis is extremely poor. The median survival for this illness from the onset of symptoms to death is less than 1 year; but there have been reported a few cases of long-term survivors[2].

Plasmacytoma and plasma cell leukemia (PCL) are both variants of plasma cell myeloma (the 2008 WHO classification) a cancer of the white blood cell responsible for producing antibodies. Both entities can present de novo, follow leukemic transformation of MM in the case of PCL or evolve into MM in the case of plasmacytoma [8,9].

A plasmacytoma is a malignant tumor of monoclonal plasma cells that can develop in either bone or soft tissue (extramedullary). Extramedullary plasmacytoma (EMP) constitutes 4% of all plasma cell tumors and it most commonly occurs in head and neck regions, its location in the peritoneum being exceptional. Other documented sites of involvement include gastrointestinal tract, CNS, urinay tract, thyroid, breast, testis, parotid gland an lymph nodes [7].
Plasma cell leukemia (PCL) is an uncommon and aggressive form of lymphoproliferative disorder characterized by a malignant proliferation of plasma cells in bone marrow and simultaneous peripheral blood involvement. The extramedullary locations of PCL are rare, as shown in a multycentric study on 73 patients with PCL, in which only one case of peritoneal involvement has been reported. Other possible but unusual extramedullary localizations are: muscular, subcutaneous, paravertebral, neuromeningeal, pleural, pericardium, pancreas and testis [4].

PCL is considered the least common variant of multiple myeloma, accounting for 2-3% of all plasma cell dyscrasias. Its prognosis is generally very poor, with a median survival of 2-8 months. But in a few individual cases of patients with VAD (infusional Vincristine, Adriamycin and Dexamethasone) regime and bone marrow/stem cell transplant there have been reported some long term survivors. Due to its rarity, there are no standard treatment regime and there is a lack of prospective data on treatment outcome in large trials for this disease [7,12-15].
Findings and procedure details

Anatomy and ultrasonographic appearance of normal peritoneum:

Familiarity with the anatomy and pathophysiology of peritoneal disease is the basis of successful ultrasound (US) study of the peritoneum.

The peritoneum is represented by a serous membrane lined with epithelial cells and divided into a parietal and a visceral sheet. The visceral peritoneum covers some of the abdominal organs while the parietal peritoneum covers the anterior and posterior abdominal walls and the pelvic cavity. The two layers enclose a large potential space, the peritoneal cavity, that normally contains up to 50-75 ml of clear fluid. The pathological processes that develop in the peritoneal cavity can disseminate through this space by the unrestricted movement of fluid and cells.

The normal parietal peritoneum appears on the ultrasound as a single, thin, smooth echogenic line in the deepest layer of the abdominal wall. This aspect is most often the combination between the peritoneum and deep abdominal fascia. If there is extraperitoneal fat in excess, the two lines appear separately. The visceral peritoneum is not usually noticeable on ultrasound in normal state but it becomes visible as a separate entity in different diseases. Bowel loops can be seen moving with respiration, independent from the peritoneum. Accumulation of fluid, like ascites, facilitates the visualization of the peritoneum and the evaluation of its pathology [10-11, 16-17].

Ultrasonographic patterns found in our patients:

The patterns that can be evaluated through US in case of peritoneal disease are:

- Ascites;
- Thickening of peritoneal reflections (including abdominal organ's capsule proliferation);
- Peritoneal nodules, plaques/sheets of soft-tissue that can progress to large masses;
- Thickening, stranding and distortion of the mesentery;
- Stranding and thickening of the omentum (omentum cake);
- Thickening and nodularity of bowell wall.

The three enteties that we evaluated have shown an US pattern of peritoneal involvement, that includes: ascites, thickening of peritoneal reflections, peritoneal nodules and plaques/ sheets of soft tissue that can progress to large masses and/or viscera invasion. The sonograms used the standard technique for abdomino-pelvic evaluation.
In the case of mesothelioma we examined a 53 year old male patient, that had previously suffered a surgical peritoneal resection for the same disease 2 years back - so this is a long term survivor (the average surviving duration for this disease being less than 1 year) with a case of relapse of the peritoneal mesothelioma, despite continuous treatment.

His US revealed:

• Parietal and visceral peritoneum diffusely thickened and involved by tumor nodules that aggregate in layers or plaques (Fig 1-4);
• No ascites, due to intense chemotherapy (ascites is a common finding in peritoneal mesothelioma - up to 90%);
• No sign of visceral invasion or capsular/subcapsular tumor masses, even though this situation has been reported as being frequent in case of peritoneal mesothelioma. Most probably, its evolution towards involvement of viscera was slowed down or prevented, due to the rapid establishment of treatment (surgery and cancer therapy), shortly after the discovery of the disease.

In the case of plasmacytoma we examined a 43 year old male patient, previously diagnosed with and treated for a vocal cords tumor and cancer of the nasal fossa, that had a history of general abdominal pain.

His US revealed:

• Peritoneal nodules originated on the anterior peritoneum, with hypoechoic appearance, irregular contours and Doppler signal, that can progress to large masses of soft tissue (Fig 5,6);
• Diffuse thickening and nodularity of parietal peritoneum - lesions of the parietal peritoneum will not move with gravity or breathing maneuvers, whereas lesions of the visceral peritoneum, mesentery, or omentum usually will move on pressure by the transducer. (Fig 6);
• Abdominal viscera, liver (Fig 7,8,9) and spleen (Fig 10), with signs of invasion, with hypoechoic, slightly heterogeneous tumor masses, located subcapsular or very close in the vicinity of the capsule.

In the case of PCL we examined a 48 year old female patient that had previously been diagnosed with acute PCL based on the hematologic profile, but was claiming abdominal fullness and diffuse abdominal pain, especially in the lower abdomen. Almost 3 months back she underwent another ultrasound that showed no collections or free peritoneal fluid, no visible nodular ultrasound images in the solid abdominal organs, and only a small fluid collection in both pleural cavities.
Her US revealed:

- Pleural effusion in both right and left pleural cavities;
- Multiple intense hypoechoic, almost transonic images, located in the middle and lower parts of the spleen, subcapsulary - possible cysts / visceral peritoneal proliferation (fig 11-14);
- Thickening of peritoneal reflections, including abdominal and pelvic organ’s capsule proliferation - spleen (fig 12) and uterus (fig 18);
- Thickening of the parietal peritoneum up to 8-9 mm, with a proliferative appearance, having irregular contours and a poor Doppler vascular signal (fig 15-17);
- Ascites in big quantity - free heterogeneous fluid in the pelvis up to 7 cm (fig 15-17), perihepatic and perisplenic fluid up to 2 - 2,5 cm, and fluid along the intestines of up to 4 cm;
- Distended bowel loops with diameters up to 4 cm, correlated with aerocolia visible on the abdominal radiography.

Two days after, the patient was brought back for another US, this time for an optimal location for paracentesis. Another few days after the paracentesis, the ascites volume continued to grow, reaching and surpassing the previous values, and the patient’s condition becoming worse, despite aggressive treatment.
Images for this section:

**Fig. 1:** Space replacement process centred on the peritoneum (fig 1,2), hypoechoic, heterogeneous, with an expansive pattern - having intense lobulated contours, a central vascular axis (with craniocaudal flow), and a thickness of about 2.3 cm.

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Fig. 2: Superficial soft tissue mass centered on the peritoneum (fig 1,2), hypoechogenic, heterogeneous, with an expansive pattern - having intense lobulated contour, a central vascular axis (with craniocaudal flow), and a thickness of about 2,3 cm.

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Fig. 3: Same patient diagnosed with mesothelioma (fig 3,4): ultrasound showing peritoneal thickening, some with deeper infiltrative growth.

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Fig. 4: Same patient diagnosed with mesothelioma (fig 3,4): ultrasound showing peritoneal thickening, some with deeper infiltrative growth.

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**Fig. 5:** Patient diagnosed with plasmacytoma. In iliac fossa: superficial large nodule (9 / 4.5 cm) with slightly irregular contours, well-defined in depth, with an uncertain interface with the anterior peritoneum. The node is well vascularized - lesion doesn't come into direct contact with the iliac vascular package.

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Fig. 6: Superficial soft tissue mass (space occupying lesion), hypoechoic, with intense irregular contours, cranio-caudal extension, mainly in surface, and a thickness of about 2 cm. The lesion seems to grow centered on the anterior peritoneum. Deeper is visualized a similar hypoechoic node, with partially erased outlines, nearly 1.8 cm in size.

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**Fig. 7:** Similar hypoechogetic lesions as those in fig 5 and 6, localized in the liver (fig 7,8,9); they are heterogeneous, with blurred contours. With the increase in dimension, the lesions become more heterogeneous and their contours more vague.

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Fig. 9: Similar hypoechoic lesions as those in fig 5 and 6, localized in the liver (fig 7, 8, 9); they are heterogeneous, with blurred contours. With the increase in dimension, the lesions become more heterogeneous and their contours more vague.

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**Fig. 10:** Similar hypoechogenic lesions as those in fig 5 and 6, localized in the spleen; they are heterogeneous, with blurred contours. With the increase in dimension, the lesions become more heterogeneous and their contours more vague.

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**Fig. 11:** Cystic lesion in the middle third part of the spleen.

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Fig. 12: In the lower ½ of the spleen - irregular microlobulated splenic contour (deposit appearance / capsular proliferation appearance)

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**Fig. 13:** Inferior pole of the spleen(fig 13,14) - cystic lesions without Doppler signal inside.

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Fig. 14: Inferior pole of the spleen(fig 13,14) - cystic lesions without Doppler signal inside.

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Fig. 15: US of the pelvis (fig 15,16,17) - ascites in large amounts, with nonhomogeneous appearance. Also at this level - peritoneal thickening up to 8-9 mm, with microlobulated contours, sometimes with almost cystic appearance (similar to those located in and on the spleen).

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**Fig. 16:** US of the pelvis (fig 15,16,17) - ascites in large amounts, with nonhomogeneous appearance. Also at this level - peritoneal thickening up to 8-9 mm, with microlobulated contours, sometimes with almost cystic appearance (similar to those located in and on the spleen); the lesions have weak Doppler signal inside.

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**Fig. 17:** US of the pelvis (fig 15,16,17) - ascites in large amounts, with nonhomogeneous appearance. Also at this level - peritoneal thickening up to 8-9 mm, with microlobulated contours, sometimes with almost cystic appearance (similar to those located in and on the spleen).

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Fig. 18: Similar lesion as those in fig 15,16,17 - peritoneal thickening with microlobulated contours, located on the anterior slope of the corporeal region of uterus.

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Fig. 8: Similar hypoechogenic lesions as those in fig 5 and 6, localized in the liver (fig 7,8,9); they are heterogeneous, with blurred contours. With the increase in dimension, the lesions become more heterogeneous and their contours more vague.

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Conclusion

Although it is not the classical imaging choice for assessing these pathologies, US is an extremely accurate imaging technique for investigation of many peritoneal pathological processes (most of them being superficial thus US provides better than CT morphological descriptions of thin lesions), also having the advantages of being a safe, relatively inexpensive and readily accessible method.

US doesn't only provide sensitive quantitative information about ascites, but also enables localization and characterization of the fluid, in which regard it is superior to CT. This information may be used in clinical practice and in research to categorize patients before beginning therapy and to assess response to treatment. In addition, US plays an important role in guidance of diagnostic and therapeutic paracentesis and allows monitoring of ascitic volume following therapeutic intervention.

The US findings may be nonspecific and not sufficient to establish a final diagnosis, but there are very useful for the detection, characterization, and guiding biopsy of peritoneal masses and also for the qualitative and quantitative informations concerning ascites - thus offering a first step in the process of diagnosis the peritoneal diseases.
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