Primary Mature Cystic Teratoma of the Liver: Report of a Rare Case

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Abstract

Teratomas are nonseminomatous germ cell tumors that arise from abnormal development of pluripotent and embryonal germ cells.[1] They usually occur in male and female gonads and are rarely reported in extragonadal sites.[2] Most findings are incidental, with imaging characteristics well described in radiological literature. To our knowledge, there have been only a few well documented cases of primary teratomas reported in the liver. Of the 26 reported, only six have occurred in adults. The majority of these cases were in female children below the age of three, mostly arising in the right lobe of the liver.[1,3,4]

We report a rare case of a mature cystic teratoma in the right lobe of the liver found incidentally in a 57-year-old female while undergoing CT scan of the abdomen and pelvis for abdominal pain related to a rectus hematoma. The diagnosis, pathogenesis, and the potential of malignant degeneration are discussed.

Introduction

Teratomas are congenital neoplasms that contain either mature or immature tissue from all three germ cell layers. Teratomas in extragonadal locations are considered very rare occurrences, with less than 1% reported to occur in the liver.[3] During fetal development germ cells follow a midline path along the urogenital ridge and descend into the pelvis as ovarian or testicular cells, hence their common midline and paramedian locations. During the first week of life, the failure of germ cells to migrate along this path and into the pelvis leads to teratomas developing in extragonadal locations.[3,5] Teratomas have distinctive imaging characteristics that allow for their easy identification, but histological examination is considered the current standard for definitive diagnosis and assessment of malignant potential.

Case Report

A 57-year-old obese female presents to the emergency room after experiencing acute onset of left-sided abdominal pain that developed 18 hours after an enoxaparin injection. Prior to her presentation, enoxaparin injections were started for anticoagulation as a bridge for a thyroid biopsy that was taking place later that week for a suspicious nodule. Her past medical history is significant for St. Jude's mitral valve replacement, long-term coumadin use, left nephrectomy for chronic hydronephrosis, cholecystectomy, hypertension, hypothyroidism, dyslipidemia, and diverticulosis. Her medical history was unremarkable for cancer.

On physical examination she was afebrile with mild hypertension. Her abdomen was obese, soft, non-distended, with fullness and tenderness to palpation in the left abdominal wall. A working diagnosis of rectus hematoma was made, and she subsequently underwent CT scan of the abdomen and pelvis with oral and intravenous contrast to evaluate the left-sided abdominal mass.

The CT scan demonstrated a 9 cm diameter hematoma in relation to the left rectus muscle. An incidental finding of a well-defined lesion within the posterior segment of the right lobe of the liver was noted to be 4.5 x 3.0cm in transverse and AP dimensions, respectively. The lesion had a thin peripheral rim.

Fatty components were appreciated, and several areas of dense material noted to be 1000 Hounsfield units (HU) with the appearance consistent to that of teeth also noted (Figure 1, Figure 2). A diagnosis of primary dermoid in the liver was made. She was admitted to the hospital at this time for treatment of her rectus hematoma and low blood count and was discharged from the hospital in stable condition. No further workup of the liver lesion was performed at this time.

Figure 1: Axial view demonstrating well-defined liver dermoid with several densities consistent to that of teeth.

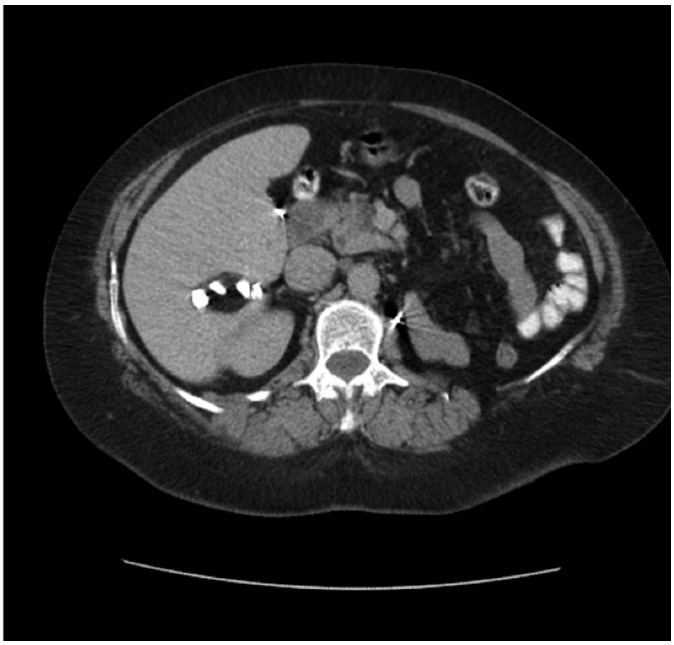
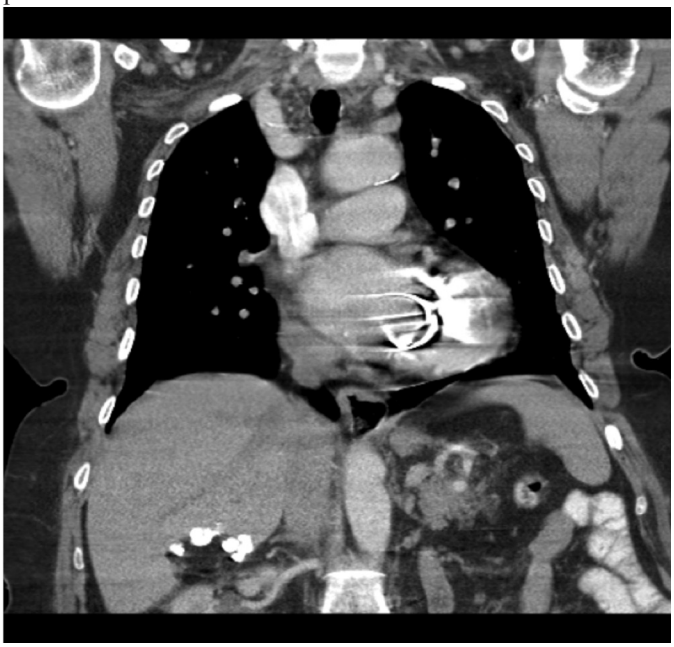


Figure 2: Coronal reformatted image demonstrating fatty components and several teeth.



A few months later she presented to her OB/GYN doctor with post-menopausal bleeding that prompted a biopsy, which revealed endometroid adenocarcinoma with squamous metaplasia. Shortly thereafter she underwent robot-assisted hysterectomy with bilateral salpingo-oophorectomy and pelvic and paraaortic lymphadenectomy. Pathology revealed lymph nodes that were negative for metastatic disease, and her final diagnosis was endometroid adenocarcinoma with focal squamous change, nuclear grade 2, FIGO 1. Approximately four months later the patient underwent follow-up CT scan of the chest, abdomen, and pelvis for evaluation of metastatic disease. Imaging revealed no metastasis and did demonstrate the same liver dermoid stable in size and appearance with no other lesions appreciated.

Discussion

Teratomas are congenital anomalies that arise from abnormal development of pluripotent cells of embryonic rests and may contain virtually any tissue type that is not typically native to the organ in which they are found.[6] Dermoid cysts and mature teratomas are synonymous, and their names are commonly used interchangeably. Dermoid cysts are considered a form of mature teratomas that predominantly contain tissue of ectodermal derivation.[1,3] They are characteristically uniloculated cysts lined by skin, and they may contain sebaceous material, hair, teeth, calcifications, or cartilage.[1]

Dermoid cysts most commonly occur in the ovary or testes with extragonadal sites reported to occur in order of decreasing frequency from the anterior mediastinum, retroperitenoum, sacrococcygeal region, central nervous system, and liver.[1] Three proposed theories exist for the causes of extragonadal teratomas. The first theory, which is most commonly cited in the literature, suggests that primary dermoids originate from displaced germ cells that arrested along the migration path from the allantois hingut to the gonads during the first week of life. The second theory suggests that they develop from supernumerary ovaries. Lastly, they are theorized to be secondary to auto-implantation of an ovarian dermoid and reimplantation of it into an extragonadal site.[7]

There are four histological variants of teratomas that are described: mature teratoma, immature teratoma, teratoma with malignant transformation, and monodermal teratoma.[1,2] Mature teratomas are usually cystic and benign but have the potential to undergo malignant transformation. This has been reported in the literature to occur in 1-2% of cases, usually in postmenopausal women during the sixth and seventh decade of life with similar imaging appearances to that of a benign mature cystic teratoma. Almost any component may become malignant, but squamous cell carcinoma accounts for 80% of cases. Complications such as cyst rupture and malignant degeneration can cause leakage of liquefied sebaceous contents into the peritoneum and further complicate the patient's course and prognosis.[1,2,8,9] Immature teratomas are much less common and usually occur during the first two decades of life. Histologically they have the presence of immature tissue. At initial presentation they are generally larger at 14-25 cm versus that of mature teratomas, which usually average about 7 cm. Immature teratomas can undergo what is known as "retroconversion" in which the tissue

can appear more mature on imaging and remain stable for long periods of time.[9] Morphologically, mature teratomas are cystic, whereas immature teratomas are predominately solid with small foci of fat.[9]

The majority of mature cystic teratomas are asymptomatic unless obstructive symptoms develop and, hence, are usually found incidentally on CT scan. Findings of a mass containing fat, fluid, and calcifications are virtually diagnostic of a teratoma.[9,10] Fat is reported in 93% of cases, with teeth or calcifications reported in 56% of cases.[9] CT scans give more specific information on fat, proteinaceous fluid, and calcifications by using HU.[11] CT scan readily identifies fat as hypodense with HU of -60 to -100, demonstrating a density similar to subcutaneous fat, and calcifications appear as markedly hyperattenuating foci.[1,3,5] Further imaging with MRI does allow for improved soft tissue identification and can be useful for diagnosing invasion of blood vessels, malignant potential, and the potential for resectability.[2,11] Mature cystic teratomas grow slowly at an average rate of 1.8 mm each year, prompting some to advocate non-surgical management of smaller, less than 6.0 cm, tumors.[9] However, definitive diagnosis is achieved by histological examination only, and surgical resection is considered the mainstay of treatment, because the presence of immature tissue affects the prognosis of the patient adversely.[2,5,6,12]

In conclusion, the case we describe here represents a rare case of primary mature teratoma in the right lobe of the liver in an adult. Although they are typically benign, malignant transformation has been known to occur and changes the patient's prognosis drastically. Our patient's repeat imaging showed the teratoma remained stable; however, the question remains as to whether this is truly a benign teratoma or somehow related to her diagnosis of endometrial cancer. Recommendations for surgical excision of the teratoma in this patient should be discussed. Although CT scans allow for the diagnosis of teratoma prior to surgery, further interventions to obtain histopathology should be considered.

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Potential Financial Conflicts of Interest: By AJCM® policy, all authors are required to disclose any and all commercial, financial, and other relationships in any way related to the subject of this article that might create any potential conflict of interest. The authors have stated that no such relationships exist.

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