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Peculiar pathognomonic ultrasound presentation of Benign cystic teratoma of the ovary: The "akara" sign

AA Adeniyi[1], E Enabudoso[2], CC Achebe[3] and SO Ukperi[3]

Department of Obstetrics and Gynaecology[1], Afe Babalola University, Ado Ekiti, Department of Obstetrics and Gynaecology[2], University of Benin, Benin City and Department of Radiology[3], Afe Babalola University, Ado Ekiti, Nigeria.

Abstract

***Introduction*:** Many sonographic features of the matured cystic teratoma have been described in literature with some such as "dermoid plugs" and Rokitansky sign being well known while others such as the "floating globules" despite being pathognomonic is less common and thus often not recognized. Because most cases of floating globules have been reported from the Europe and Asia, the nomenclatures or names given to this finding have been related to events in these regions. Thus, terms such as "truffles sign", "poke ball sign" have been used to described cases of floating globules, while these terms are familiar to the European and Western populations, they are undoubtedly alien to the African culture.

Methods: This is a case report of a 40-year-old woman who presented with classical ultrasound features of floating globules. To the best of our knowledge no ultrasound features similar to the "floating globules" of benign cystic teratoma have been reported from Africa. Our case described the sonographic findings of floating globules in a Nigerian woman. We also proposed a familiar name "akara sign" for this intriguing and peculiar feature of benign teratoma.

Conclusion: The case highlighted the ultrasound findings of floating globules in an African woman and also proposed a familiar name for the phenomenon for easier recognition.

Keywords: Benign cystic teratoma, floating globules, ultrasound scan, "akara" sign

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Correspondence: Dr. A.A. Adeniyi, Department of Obstetrics and Gynaecology, Afe Babalola University, Ado Ekiti, Nigeria. E-mail: adeniyiaa@abuad.edu.ng

Introduction

Matured cystic teratomas of the ovaries are regarded as the commonest benign ovarian tumors [1,2]. They are a variant of germ cell tumors of the ovary and they arise from two or more of the three primordial body layers; endoderm, mesoderm or ectoderm [2]. The commonest form of these tumors is the dermoid cysts which arise mostly from the ectodermal layer and constitutes about 70% of teratomas [3]. Many classical sonographic features of the matured teratoma have been well described, such as "dermoid plug" also known as Rokitansky noddle or protuberance, which is a mural hyperechoic mass, ''the tip of the iceberg sign'', an echogenic interface at the edge of mass with posterior shadowing. Others are "dot-dash pattern" or "dermoid mesh", which are

multiple thin echogenic strands caused by the presence of hair in the cyst cavity [2]. A rarer but pathognomonic sonographic feature of the matured teratoma has also been reported mostly from Europe and Asia [4,5]. It is the mobile globules or floating balls in benign cystic teratoma of the ovary. To the best of our knowledge no such case has been reported from African population. Thus, we present the sonographic findings of such case in a Nigerian woman, while also proposing a name that is culturally more understandable in our environment- the "akara" sign.

Case presentation

A 40-year-old multiparous woman who present as a case of secondary infertility of 5 years duration at our facility. There was no significant medical or surgical history. She was married to 43-year-old man. She neither smoked cigarettes nor drank alcohol. Findings on general physical examinations were essentially normal. Examination of the systems were also normal except for a non-tender suprapubic fullness. Pelvic examination revealed a normal sized uterus which was freely mobile with no associated cervical excitation tenderness. The left adnexum appeared full but non-tender. Pelvic ultrasound scan revealed a normal sized uterus with uniform myometrium. The right adnexum was normal. There was a left ovarian cystic mass with multiple rounded discrete globules floating within a cystic background. Each globule measured between 2cm to 4cm in diameter and were not attached to each other or to the wall of the cyst (Figure 1). The findings were found to be identical with the previously described, rare but pathognomonic feature of benign cystic teratoma of the ovary.

Discussion

Mature cystic teratomas (MCT) also known as "dermoid cyst" have been described as one of the commonest germ cell neoplasms of the ovary [6,7]. They accounted for about 10 -25% of ovarian tumours in adults and up to 50% in younger age group [8]. Ultrasound provides an easy and reliable technology for their diagnosis. Many sonographic features have been described. Many of these ultrasound findings occur with predictable consistency such as "dermoid plugs" or Rokitansky noddle, tip of the iceberg sign, dot-dash sign, comet tail appearance, fat-fluid, fluid-fluid level and dermoid mesh [2,9,10].

A particular rare but pathognomonic sonographic feature of benign cystic teratoma is the floating globules which are discrete rounded masses measuring an average of 2-4cm in diameter within a cystic background [2,4,5](Fig.1). The pathogenesis of these floating globules has not been clearly elucidated, but each globule is believed to have been formed by the aggregation of sebaceous matter around a focus of debris or hair shaft [4]. Different names and nomenclatures have been used to described this phenomenon depending on the country of the author(s). These names usually compare the sonographic finding with a familiar object or phenomenon in the locality of the author.

Such names include; "truffle sign" because of its similarity to chocolate truffles [4], "poke ball" sign in relation to Pokémon balls [5]. These names are quite alien to most African population where to the best of our knowledge no such case has been reported. We believe that it is better to describe this rare but pathognomonic sonographic feature of benign cystic teratoma with a name that is familiar

Figure 1. Sonographic image of the left Ovary of the Patient showing the floating globules

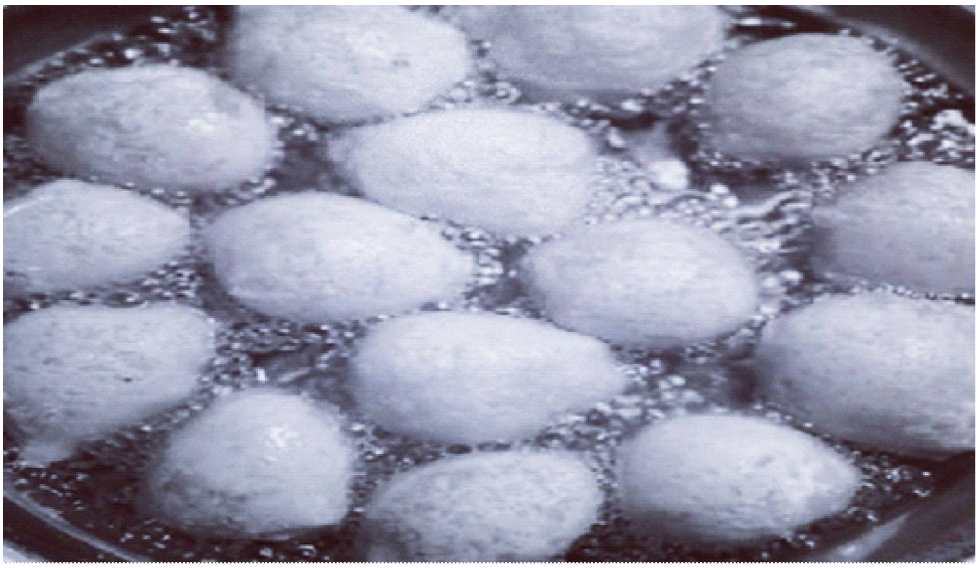

Figure 2: Picture of "akara" in a frying pan (similar to the floating globules in cystic teratoma)

Figure 3: Floating globules '' Truffle sign'' in Matured cystic teratoma as reported by Otigbah et al[4]

to the population where this case is being reported. This will facilitate easy recognition and diagnosis. The sonographic image of floating globules discussed in this case bears a close resemblance to a delicacy eaten in most African countries and known by a common name in these countries as "akara" (or beans cake) in fry (Fig.2) Thus, we proposed the name "akara sign" (or beans cake sign) to describe the phenomenon of floating globules in benign cystic teratoma. It is also noted from previous studies that this pathognomonic sonographic feature is also consisted with the histological findings of benign teratoma [4,5] and thus the use of more expensive imaging modalities such as magnetic resonance imaging (MRI) or computed tomography (CT) may not be necessary for diagnosis of this condition once the classical ultrasonic features as reported in this case and as reported by Otigbah et al (figure 3) and other Authors [2,4,5] are present. This is especially important in low-income settings where these techniques are either not available or affordable and requesting for such investigations will only lead to delay in diagnosis and treatment.

The major limitation of the case presented is the fact that data were not available on the clinical and surgical outcomes of the case as the patient defaulted from further management and follow up. However, the sonographic features presented will help in quick identification and diagnosis of such cases in the future.

Conclusion

While the pathognomonic sonographic feature of floating globules in matured teratoma have received many names, we proposed to add a familiar name; "akara sign" to the list for better recognition and easy diagnosis especially in African population where the feature bears a close semblance to "akara" a common delicacy.

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