JUVENILE CYSTIC ADENOMYOMA- A DIAGNOSTIC ENIGMA

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ABSTRACT:

Juvenile cystic Adenomyoma(JCA) is a rare form of adenomyosis, affecting young girls. We are presenting three cases of JCA misdiagnosed preoperatively, as unicornuate uterus with haematometra in non communicating rudimentary horn in two cases and fibroid with cystic degeneration in one case. MRI pelvis is the investigation of choice. Complete excision of the lesion done in all the three cases with recurrence in one case.

KEYWORDS: Adenomyoma, Dysmenorrhea, Mullerian Anomalies, Juvenile Cystic Adenomyoma.

Introduction:

The presence of heterotropic endometrial gland in the uterine myometrium with smooth muscle hyperplasia is called adenomyosis.It is usually seen after the age of 30years.[1] The diffuse variety occurs more commonly which is characterized by cystic spaces of <5mm diameter filled with blood. While, the cystic variant called “adenomyotic cysts” or “ adenomyomas”[6] is extremely rare especially in paediatric population.We are presenting 3 cases of JCA.

Case1 :

A 20year old lady, married, nulliparous, presented with progressive dysmenorrhea for 2years. Pain was spasmodic in nature, non-radiating, associated with vomiting, giddiness and syncopal attacks and was refractory to analgesics.Trans abdominal scan showed a normal sized uterus with rudimentary horn with hematometra (3.5cmx4cm). On performing mini-laparotomy, uterus size was normal with a bulge seen in the right anterior wall below the origin of the round ligament of size 3x4cms. Vasopressin was injected and complete excision of the cyst was performed and the dead space was obliterated with 1-0 vicryl suture. On cut section thick chocolate colored fluid was drained. Histopathalogical analysis confirmed the diagnosis of adenomyotic cyst. Her pelvic pain and dysmenorrhea were completely relieved after surgery. She is yet to conceive.

Case2:

A 15year old girl presented with severe cyclical pain since 1 year, spasmodic in nature, non-radiating, not associated with any gastrointestinal and urinary symptoms refractory to analgesics and Oral contraceptive pills. Minimal tenderness present in the right lower abdomen. Trans abdominal scan showed normal sized uterus with Right rudimentary horn with hematometra (2x3cm) MRI pelvis showed Adenomyoma with altered echotexture on the right cornualend of the uterus of size 2x3x2cm. Mini laparotomy was performed, intraoperatively, uterus size was normal with a bulge seen on the right anterior wall below the origin of the round ligament of size 2x3 cms. Vasopressin was injected and wide excision was performed with myometrial closure with vicryl 1-0. On cut section thick chocolate colored fluid was drained. Histopathalogical analysis confirmed the diagnosis of adenomyotic cyst. After 6 months patient came back to us with similar complaints. Review Transabdominal scan showed Adenomyotic cyst with collection of size 1x2cms sign of recurrence. She was treated with Tab. Dienogest 2mg once a day for 3 months and was asked to report with Surgery SOS.

Case3:

A 18year old girl presented with progressive dysmenorrhea, spasmodic in nature, non-radiating associated with vomiting and involuntary movements like head banging for which psychiatry consultation was done and was diagnosed to have PremenstrualDysphoric disorder with behavioural changes and was treated accordingly. Pain was refractory to analgesics. She attained menarche at 12 years of age. She had regular cycles with average flow. No tenderness was noticed on examination of the abdomen. Trans abdominal scan showed normal uterus with evidence of well-definedheterogenouslyhyperechoic lesion of size 27x25mm in the fundal region with no significant vascularity likely Fibroid. MRI pelvis showed Uterine fibroid with central cystic degeneration (33x30mm). Mini-Laparatomy was performed;intraoperatively uterus was normal in size with fibrotic myometrial lesion of 4x3cms noted just below the attachment of the round ligament on the right side. Vasopressin was infiltrated and wide excision was performed. Myometrial closure was done with vicryl 1-0. On cut section chocolate coloured fluid was drained from the cyst. Histopathalogical analysis confirmed the diagnosis of adenomyotic cyst. Her pelvic pain and dysmenorrhea were completely relieved after surgery.

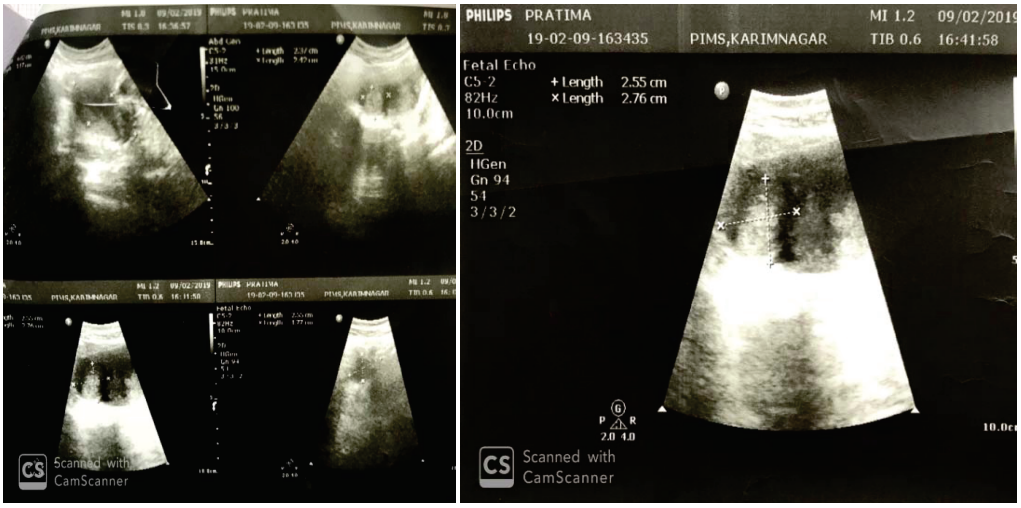

Figure 1 Transabdominal scan showing fundal fibroid of size 25x27mm.

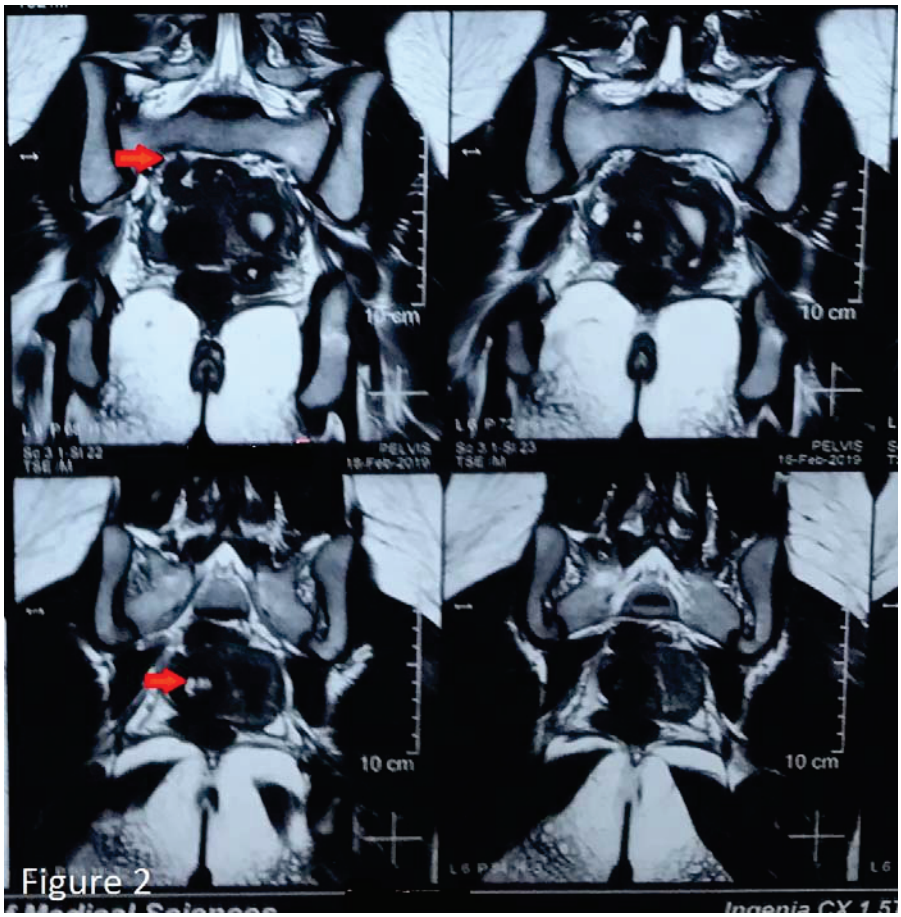

Figure 2 MRI pelvisshowing the uterine fibroid with central cystic degeneration (30*33mm) in the coronal section.

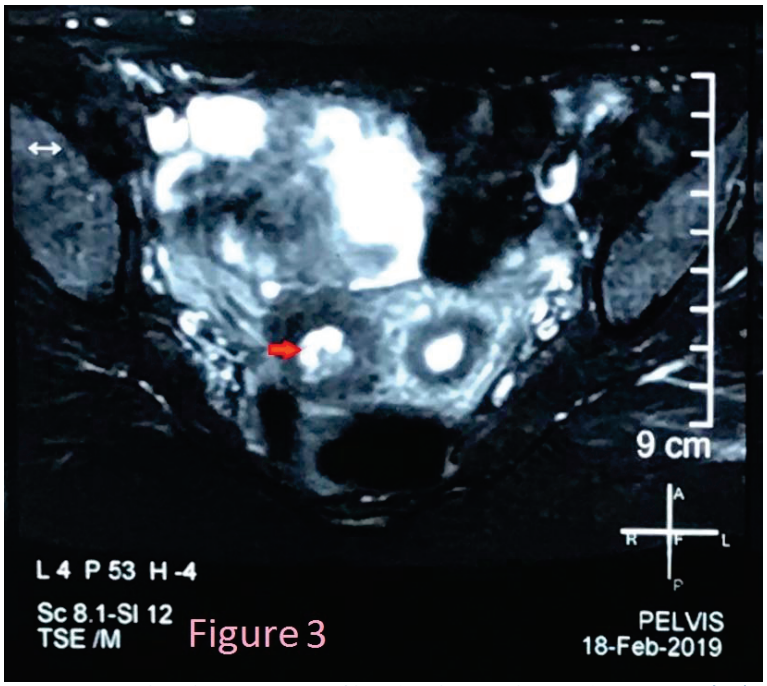

Figure 3 MRI pelvis showing uterine fibroid with central cystic degeneration(30*33mm) in the tranverse section.

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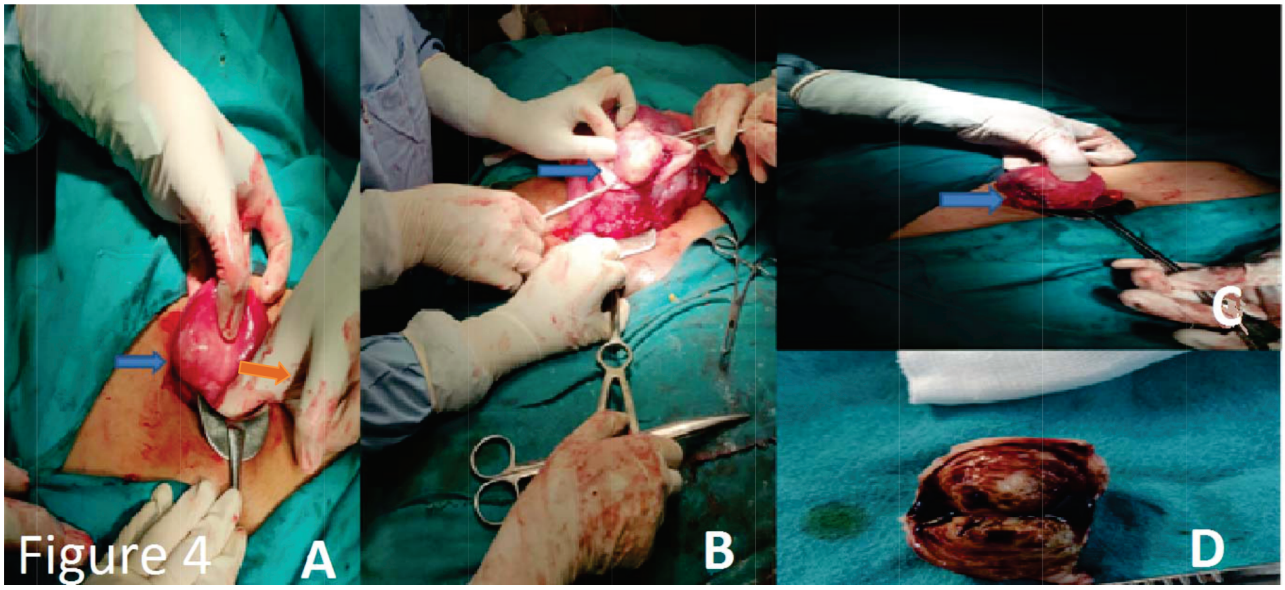

A. Normal sized uterus with bulge on right side of size 4x3 cms noted just below the attachment of the round ligament
B. Complete excision of the cyst
C. Myometrial closure done with 1-0 vicryl.
D. Cut section showing chocolate colored fluid drained out

Discussion:

The actual incidence and aetiology of JCA is unknown. Many cases of JCA are now being reported because of improved imaging techniques and increased awareness. Recently these cystic adenomyomas are categorised under Accessory and cavitated uterine masses (ACUM)[1],[3]. Because of its juvenile onset and peculiar location which is always on the anterior wall of the uterus near the origin of the round ligament,Acién et al. [1]have proposed that these type of juvenile cystic adenomyomas should be considered as a new type of congenital mullerian anomaly. These lesions can be caused by the duplication of the ductal mullerian tissue at the origin of the round ligament, which could be due to the gubernaculum dysfunction.[4] It also suggest that these cystic adenomyomas should be seperately classified from the rest of the mullerian anomalies as the uterine cavity contour is preserved.[4]

Takeuchi et al [5], have considered the juvenile cystic adenomyoma as a cystic variant of adenomyosis, rather than considering it as a congenital anomaly. A diagnostic criteria was provided by Takeuchi et al [5] ; this included:

A) Age of 30 years or younger.
B) Cystic lesion ≥ 1 cm in diameter and independent of the uterine lumen and covered by hypertrophic myometrium on imaging modality or intra operatively.
C) Associated with severe dysmenorrhea.

Dysmenorrhea may be due to the intracystic bleeding and stretching of the cystic cavity.[6]Ultrasonography is the first imaging method performed.Though MRI is the modality of choice, results were misinterpreted in our cases.[6]

In our cases we suspected juvenile cystic adenomyoma, as the uterus contour did not suggest a rudimentary horn on performing mini- laparotomy and the salient feature is the location, it is near the origin of the round ligament, all our cases had the same finding and was hence confirmed.[6]

Medical treatment withnon steroidalanti inflammatory drugs, contraceptive pills and gonadotrophin releasing hormone analogues is limited and provide only temporary relief.[4]The gold standard treatment for JCA is complete excision of the lesion.[4][5][6]

Conclusion:

Many cases reported in the literature were misdiagnosedpreoperatively as:[6]

a) Mullerian anomalies
b) Cystic degeneration ofleiomyomas, and
c) Broad ligament fibroids

Gynaecologists, as well as radiologists, should be aware of and keep a high index of suspicion about this rare condition to make a proper preoperative diagnosis.

JCA should be kept in mind as a differential diagnosis in young girls with severe dysmenorrhea as it is a treatable cause. Complete exicision of the lesion is the only definite therapeutic option.

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