Big Perineal lipomas with imperforate anus

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Abstract

Newborn male patient presented at alkhansaa teaching hospital with signs of neonatal intestinal obstruction secondary to big perineal lipoma and anorectal malformation, a colostomy performed .After 4 months the size of lipoma get bigger, a decision for excising lipoma with posterior sagittal ano-recteoplasty was performed .Regular dilatation for the internal anal sphinector and closure of colostomy was done ,follow up for 5 years shows normal bowel function with total continence t although it has been said that perineal lipoma with anorectal malformation may compromise bowel function ,we conclude that the line of management is the same as simple anorectal malformation and the outcome is promising if the exicision of lipoma done early in life.

Key ward : perineal lipoma, anorectal malformation.

Introduction

Newborn with anorectal malformation may have associated perineal lipoma ,these lesions are rare (1).

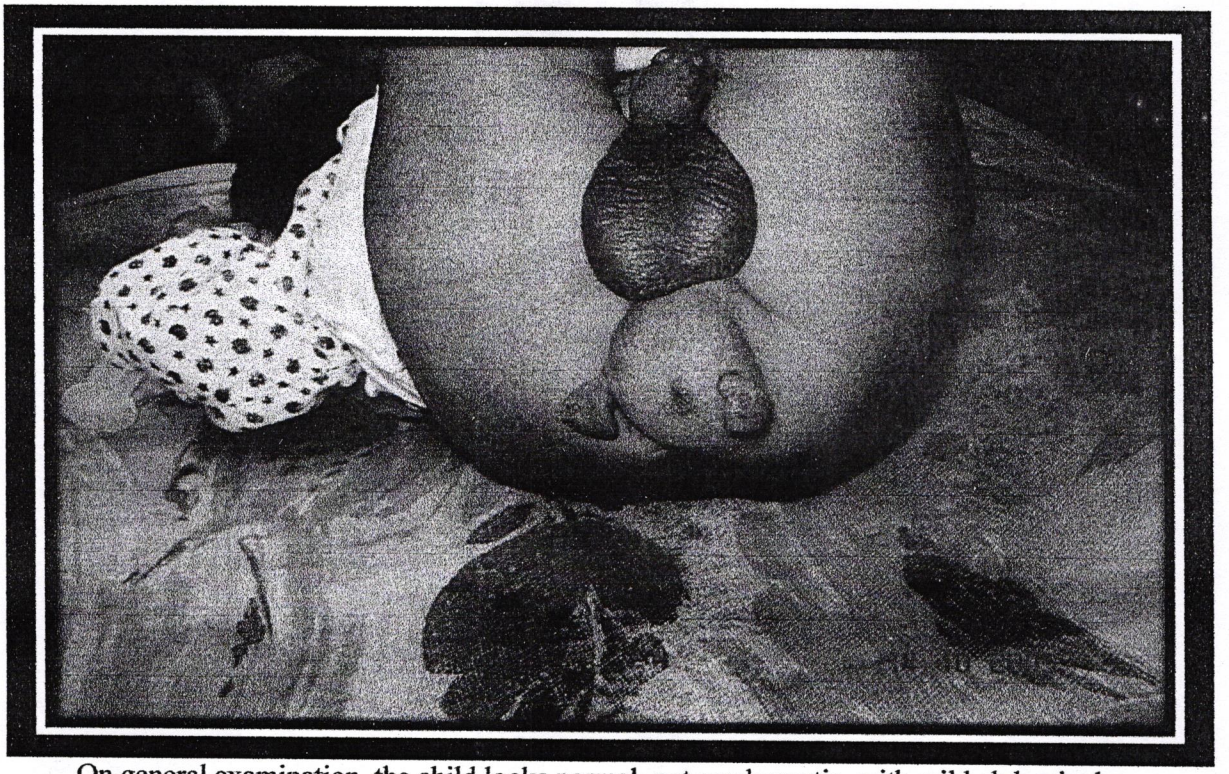
The presence of unusual perineal masses can add to the complexity of ano-rectal malformation ,regarding differential diagnoses like meningocele ,myelomeningocele,teratomas or awareness in the protocols of surgical correction (2),(3).

We describe a newborn male with ano-rectal malformation and big perineal mass with necessary diagnostic investigation to be done in order to achieve best lines of surgical procedure to be performed .which are ultrasound,, complete blood picture ,chest x-ray ,

Patient and method

An Iraqi male , born with 3100g. weight at birth, was transferred to Al-Khansaa teaching hospital, pediatric surgery center ,for evaluation and management of imperforate anus and a big perineal mass . He was the third child in the family ,with no previous history of same pathology ,the 35 years old mother neither had antinatal care nor ultrasonography but she described polyhydromina. The child was born by spontaneous vaginal delivery with normal apgar score.

DOI: http://dx.doi.org/10.25130/mjotu.18.1.17



On general examination ,the child looks normal ,not syndromatic, with mild abdominal distension . His external genitalia was normal , moving both lower limbs, and a big soft mass was noted at the midline of perineal region slightly to left . with two small tags at both side.

At 36 hours of age he passed meconieum per urethra ,kept on ampicilline and gentamycin according to his weight , vitamin k as prophylaxis .Nothing by mouth and intravenous fluid.

Antero-posterior and lateral radiographs of the sacrum and coccyx revealed normal bony structure , no calcification with a column of air distending the rectum of high type.

An ultrasound and echocardiogram were done to asses cardiac, renal anomalies ,both were negative.

Ultrasound of the mass revealed a 6*3*2 cm lesion with no interpretation about extension into the presacral space .The serum alpha-feto protein was normal for his age. MRI was not available in our center.

The patient underwent first stage surgery consisted of creation of colostomy and mucous fistula with uneventful recovery and patient discharged 3 days later.

He was kept on strict observation for any new change in mass like increasing in size , infection or getting tense for a period of 4 months.

Before definitive surgical repair ,re-evaluation of the mass was done which shows bigger size , a contrast enema of distal colon was performed to delineate the level of his rectum ,the rectal pouch not dilated ,about 5 cm from the perineal region with clear fistula to the urethra , again there is soft tissue mass with no calcification inside.

At 4 months of age with 6 kg body weight he underwent posterior sagittal ano-rectoplasty (psarp) and excision of perineal mass . A mid line incision was made anterior and posterior to the perineal mass ,and the skin of the mass was surrounded circumferentially {figure appropriate anal size for age was reached . the five years old male 2}, total excision of the mass performed and histopathology revealed benign fatty tissue {figure 3} ,the surgical field at this step like any ordinary other (psarp) , identification of recto-urethral fistula with suturing , pulling the rectum down to perineum
{figure 4} with drain due to extensive dissection .The patient recovered without complication , kept on regular dilatation until patient is now doing well with normal bowel function after colostomy closure.

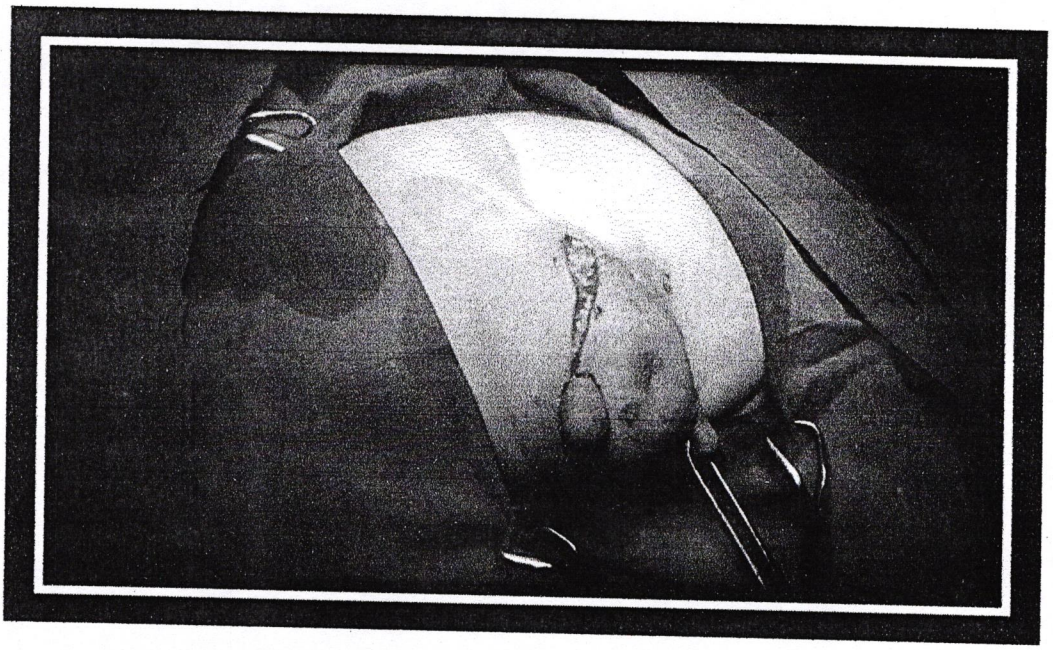


Figure 2 Mid line incision circumferential around the mass

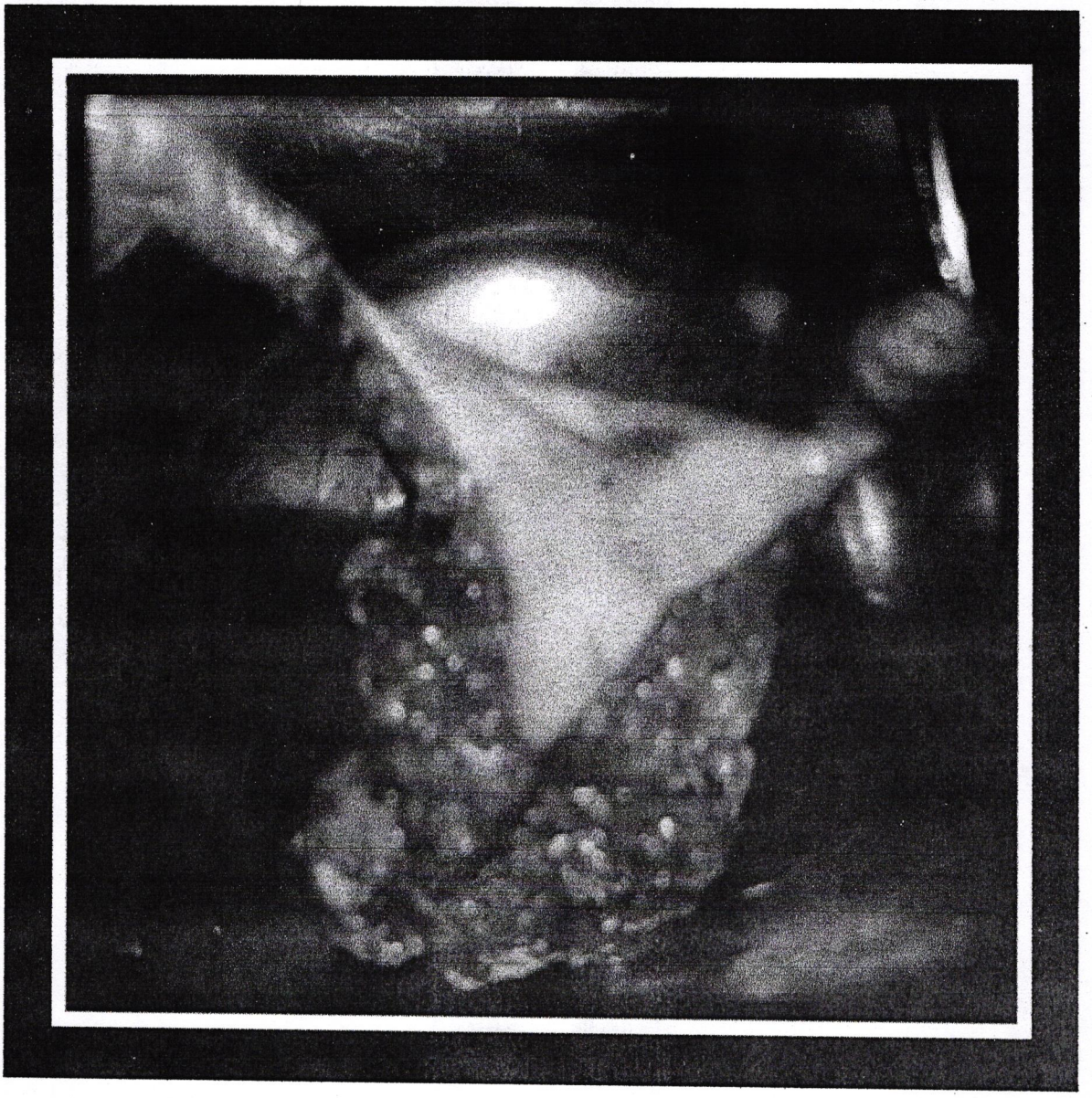


Figure 3 The mass totally excised

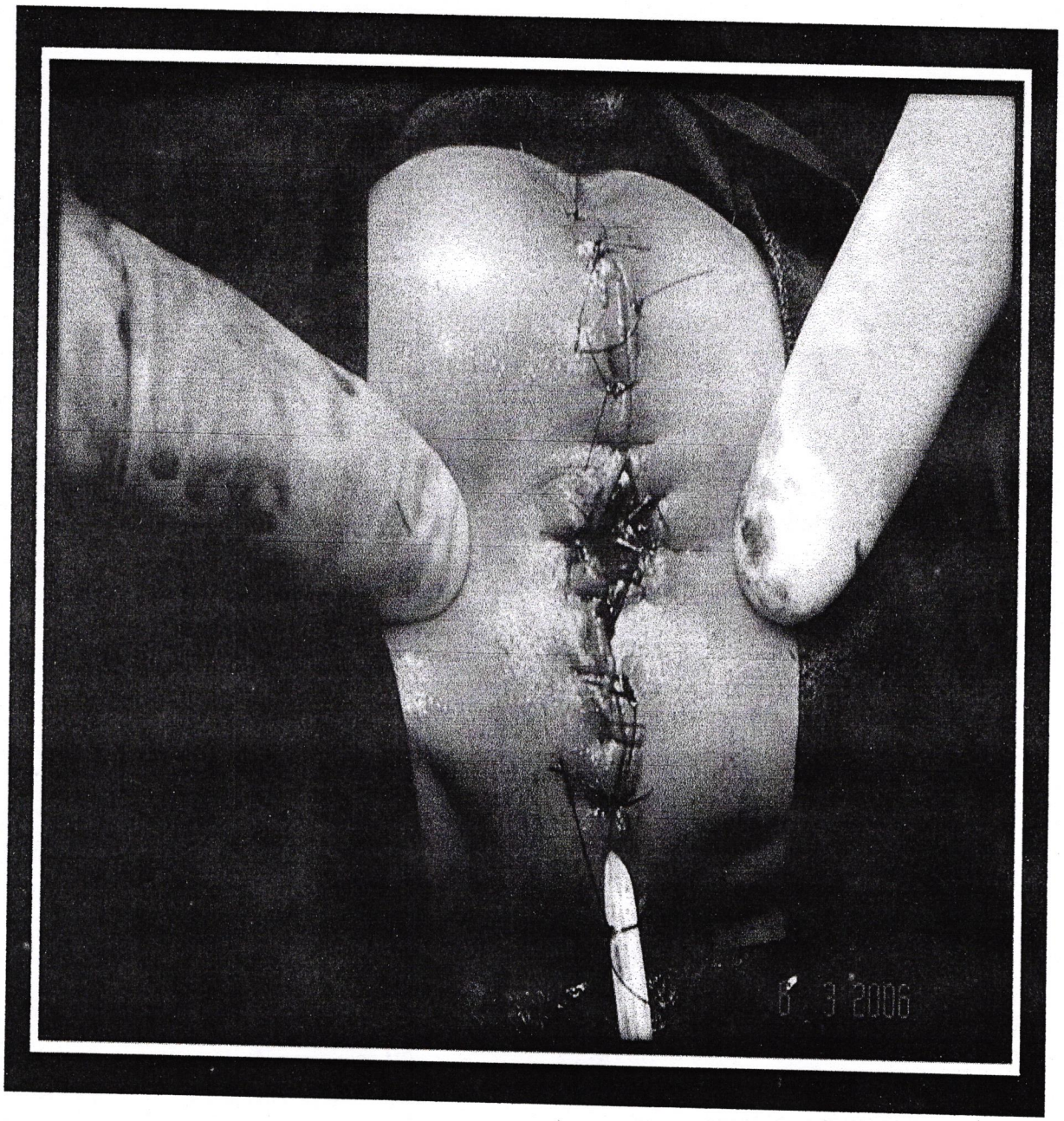


Figure 4 New anal opening with drain

Discussion

To the best of our knowledge There is no much information about association between ano-rectal malformation and big lipoma in the perineum as isolated entity and not syndromatic .3

This case report to delineate the possible and safe lines of management for such a rare pathology according to what written in published articles , in order to set a protocol about when and how treat these cases .

A true tail is defined as the remnant of embryonic tail, which usually regresses during the seventh and eight weeks of gestation, a psueudotail is a protrusion from the lumbosacrococcygeal area that may be composed of normal tissue (4).

In comparism with other case report published it seems to be that the best way of treatment is to do thorough pre operative evaluation to delineate anatomy and to list the differential diagnoses, starting with diverting colostomy, followed by excising the mass with (psarp) at the same session in short period if one suspect serious pathology ,then keep on dilatation till colostomy take down .

At least follow up period should be not less than 2 years to asses bowel function which is like to be good as in our case .

Conclusion :

Anorectal malformation with a big perineal mass association is rare , need extensive investigation and expert pediatric surgeon in these anomalies to deal with it as such. The presence of a mass does not affect the usual line of management of simple ano rectal malformation in general.

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