

Case Report

Dorsal accessory ectopic breast with polythelia – A marker of occult spinal dysraphism

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Abstract

Background: Accessory breast, also known as supernumerary breasts, polymastia, or mammae erratae, is a clinical condition of having an additional breast. Accessory breasts are usually seen along the embryonic milk line, with the majority located in the axilla. Polythelia is the presence of an additional nipple. We report a rare case of dorsal accessory ectopic breast with three nipples (two well formed and one rudimentary) occurring along with lipomeningomyelocele and diastematomyelia.

Case Description: We report the case of an 18-year-old female who presented with chief complaints of swelling over the upper back since birth and spastic weakness of bilateral lower limbs with inability to walk since 2 years. Three-dimensional computed tomography scan of the dorsal spine was suggestive of a wide bony defect in the posterior spinal elements from D3 to D9 vertebrae. Diastematomyelia was also seen. Magnetic resonance imaging of the dorsal spine was suggestive of a complex spinal dysraphism with lipomeningomyelocele and diastematomyelia. During surgery, the patient's accessory breast was removed, lipomatous tissue and bony septum were excised, and dural repair was done. Histopathological examination was consistent with accessory ectopic breast with lipomeningomyelocele.

Conclusion: Dorsal accessory breast, although a rare entity, whenever present should alert the clinician regarding the possibility of an underlying occult spinal dysraphism (OSD). Therefore, dorsal accessory breast can also be considered as a marker of OSD.

Key Words: Dorsal, ectopic breast, lipomeningomyelocele, occult spinal dysraphism, polythelia

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INTRODUCTION

Accessory breast, also known as supernumerary breasts, polymastia, or mammae erratae, is a clinical condition of having an additional breast. Accessory breasts are usually seen along the embryonic milk line, with the majority located in the axilla. Polythelia is the presence of an additional nipple. We report a rare case

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of dorsal accessory ectopic breast with three nipples (two well formed and one rudimentary) occurring along with lipomeningomyelocele and diastematomyelia. The association of dorsal accessory breast with meningocele and diastematomyelia has been reported only once in the world literature, however, the association of dorsal accessory breast with polythelia with lipomeningomyelocele and diastematomyelia has never been reported.^[4]

CASE DESCRIPTION

We report the case of an 18-year-old female who presented with chief complaints of swelling over the upper back since birth and spastic weakness of bilateral lower limbs with an inability to walk since 2 years. On examination, the swelling was soft-to-firm, nontender, midline in location, which started increasing in size at puberty and progressed since then to the present size of 14 cm × 12 cm [Figure 1]. Three-dimensional (3D) computed tomography scan of the dorsal spine was suggestive of a wide bony defect in the posterior spinal elements from D3 to D9 vertebrae. Diastematomyelia was also seen from D3 to D7 vertebrae [Figure 2].



Figure 1: (a) Clinical examination of the patient showing a midline dorsal accessory breast with 3 nipples (2 well formed and 1 rudimentary). (b) Close up view

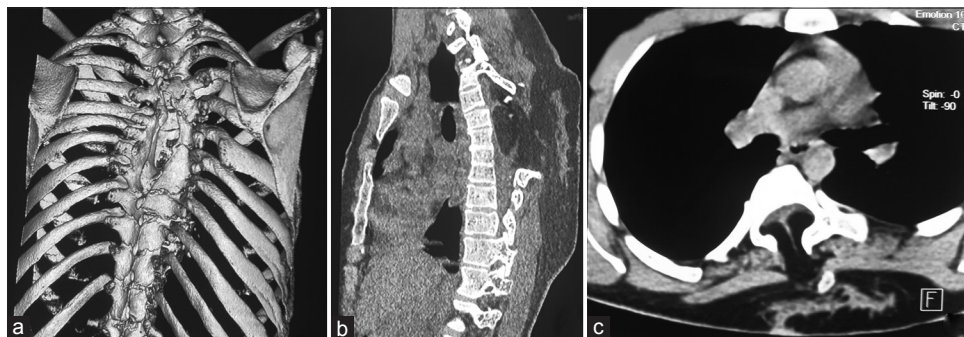


Figure 2: (a and b) 3D Computed Tomography (CT) scan of the dorsal spine showing a wide bony defect in the posterior spinal elements from D3 to D9 vertebrae. (c) Diastematomyelia is also seen

Magnetic resonance imaging (MRI) of the dorsal spine was suggestive of a complex spinal dysraphism with lipomeningomyelocele and diastematomyelia [Figure 3]. At surgery, the patient's accessory breast was removed, lipomatous tissue and bony septum were excised, and dural repair was done. An elliptical vertical skin incision was taken incorporating the reductant skin along with the three nipples. The entry of the fibrolipomatous stalk through the fascial defect was identified. A large lipomatous mass along with the accessory breast was amputated at the level of the stalk to help in the visualization of the spinal cord–lipoma interface. Two hemicords with a dorsal type lipomeningomyelocele was identified [Figure 4]. The penetrating intradural lipomatous tract was identified all around. The osseocartilaginous median septum was drilled. Normal dura of both the hemicords was opened and the openings were carried caudally towards the penetrating tract. Circumferential dissection around the lipomatous tract was continued until the dura was separated completely. The dural opening was then continued caudally. The fatty mass was resected with the help of cavitron ultrasonic surgical aspirator (CUSA). Neurulation of the neural placode was then done. Water tight dural closure (duraplasty) was done for both the hemicords separately with the help of a graft. Histopathological examination was consistent with accessory ectopic breast with lipomeningomyelocele [Figure 5].

The postoperative period was uneventful. Spasticity and power in the lower limbs improved and the patient was able to walk with support at the time of discharge.

DISCUSSION

Multiple theories have been proposed to explain the occurrence of accessory breast. These include Darwin's theory which stated that "traits which have disappeared generations before, can reappear," Pfeifer's theory of metaplasia of sweat glands or modified sweat glands, Hughes's theory of random migration of primordial breast cells away from the mammary crest, and Schultz's theory of displacement of milk lines, laterally or caudally.^[2,5-7]

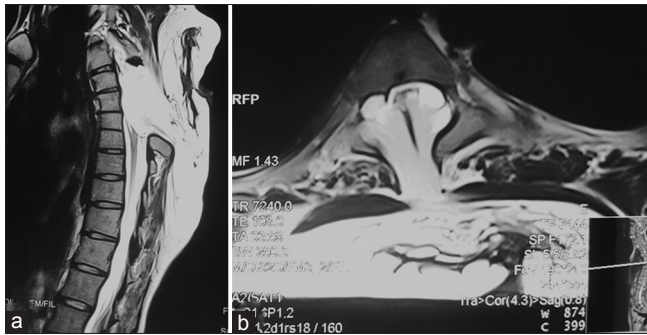


Figure 3: (a) Magnetic Resonance Imaging (MRI) of the dorsal spine suggestive of a complex spinal dysraphism with lipomeningomyelocele. (b) Diastematomyelia is also seen

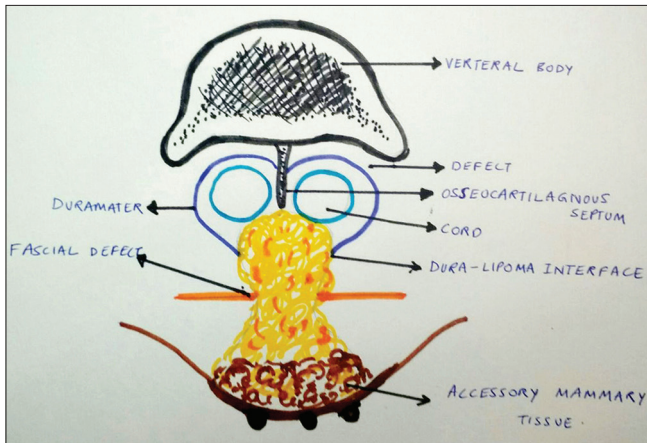


Figure 4: Schematic diagram showing the relationship of occult spinal dysraphism (OSD) with dorsal accessory ectopic breast

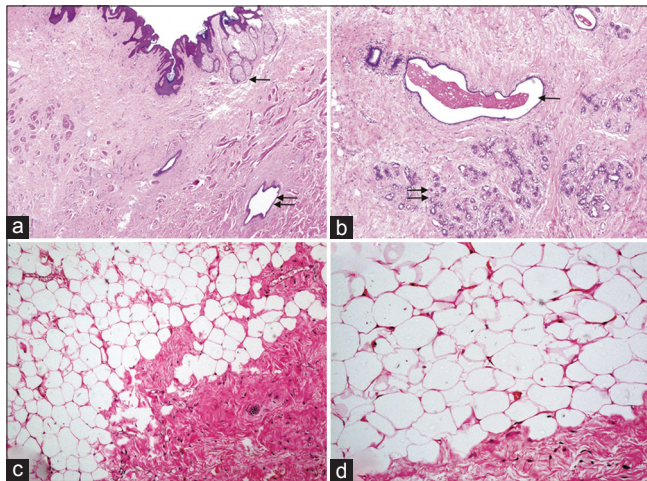


Figure 5: (a) Hematoxylin and eosin stained section (x40) showing normal skin with appendages (single arrow); Subepithelial tissue shows lactiferous duct (double arrow). (b) Hematoxylin and eosin stained section (x80) showing lactiferous duct with secretion inside (single arrow) along with terminal duct lobular units of breast (double arrow). (c and d) Hematoxylin and eosin stained section (x20 and x40 respectively) showing lipomatous component composed of sheets of mature adipocytes with adjacent dural fibroblasts

Studies have shown that an ectopic breast usually increases in size after hormonal stimulation during

puberty, pregnancy, or lactation.^[1] In our case also, the swelling started increasing in size after the girl attained puberty. Ectopic breasts, similar to normal breasts, are prone to the risk of developing various types of breast diseases such as abscess, mastitis, fibroadenoma, fibrocystic disease, or breast cancer.^[8]

Occult spinal dysraphism (OSD) is characterized by skin-covered lesions without exposed neural tissue. Cutaneous markers of OSD include lipoma, fibroma pendulum, human or faun tail, dermal sinus, atypical dimple (larger than 5 mm and more than 2.5 cm from the anus), hamartoma, aplasia cutis congenita, deviation of the gluteal furrow, port wine stain, hemangioma, hypertrichosis, and acrochordon (skin tag).^[3] A combination of two or more congenital cutaneous lesions constitutes the strongest marker of OSD.^[3]

CONCLUSION

Dorsal accessory breast, although a rare entity, whenever present should alert the clinician regarding the possibility of an underlying OSD. Therefore, dorsal accessory breast can also be considered as a marker of OSD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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