Idiopathic hemihypertrophy with multiple fibroadenoma

Sir,

Idiopathic hemihyperplasia or hemihypertrophy (HH), is an uncommmon entity reported in approximately 1:13,000 to 1:86,000 live births and it is characterized by asymmetric growth of body parts.[1]

We present a case of a 16-year-old female who came to us with complains of multiple swellings in bilateral breasts for the last one year along with a single brown patch over her right breast and body asymmetry since birth. Her parents gave history of her body being larger and heavier on right side since birth and the disparity of size was decreasing slowly with age. No history of similar illness in family, and no history of consanguinity in parents was noted.

On examination, we found hemihypertrophy on right side including face and verrucous epidermal nevus of the size 2 × 4 cm on right breast [Figure 1a and b]. Mid-arm circumference of right upper limb was 1.5 cm more, whereas midthigh and midcalf circumference was 3 cm more on right side. Also lower limb length discrepancy was there as right lower limb was 2.5 cm longer and right knee level was higher in standing position [Figure 1a and c]. There was hyperpigmentation of the epidermal nevus clearing of its own at one edge and progressing on the other edge. Multiple mobile, firm swellings were felt in both breasts. No thickening of palmar surface or macroglossia was evident. The patient's IQ and neurological examination were normal. Skull X-ray AP view showed facial disparity due to relatively smaller left orbit and left hemimandible [Figure 1d]. Ultrasonography of breast showed multiple fibroadenomas on each side of breast with largest being of size 6 × 3 × 2 cm on right side [Figure 2a]. Routine blood investigations, liver function test, kidney function tests, serum AFP level, chromosomal analysis [Figure 3a], ultrasound of abdomen and magnetic resonance imaging of brain were normal. Particular gene analysis was not done due to unavailability.

Histopathological examination of nevus tissue revealed features consistent with verrucous epidermal nevus [Figure 3b]. Aspiration cytology of one of the breast swelling was suggestive of fibroadenoma [Figure 2b]. Based on clinical, radiological, and pathological findings, a final diagnosis of idiopathic hemihypertrophy (IH) with multiple fibroadenomas and verrucous epidermal nevus was made.

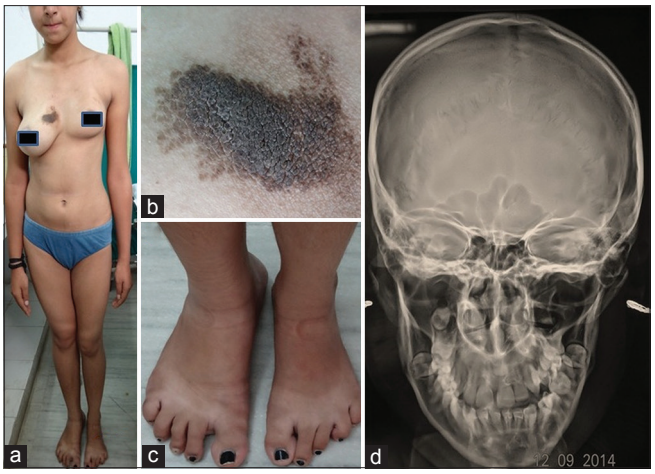

Figure 1: (a) Asymmetry of the trunk and face (hemihypertrophy), (b) verrucous epidermal nevus over breast, (c)right foot hypertrophy, (d) skull X-ray shows bony facial asymmetry

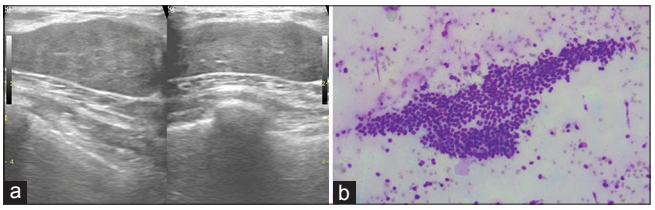

Figure 2: (a) USG right breast showing smooth margined hypoechoic fibroadenoma which is confirmed on (b) Giemsa stained cytology smear of the lesion showing cohesive clusters of ductal epithelial cells in background of bare bipolar cells

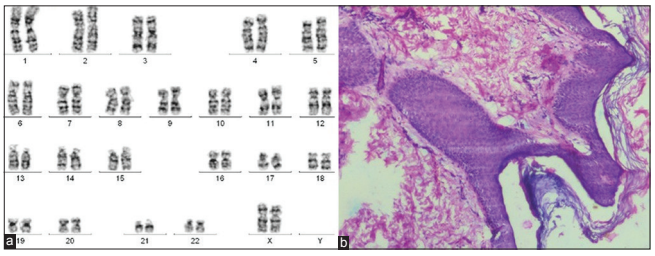

Figure 3: (a) Chromosomal analysis (GTG-Banding with 500 band resolution) showing normal female karyotype (b) Histopathological section of the skin lesion is showing hyperkeratosis, acanthosis, and papillomatosis favoring epidermal naevus

Hemihyperplasia or hemihypertrophy is an abnormality of cell proliferation leading to asymmetric overgrowth of one or more body regions. Hemihypertrophy can occur as an isolated abnormality or as a component of overgrowth syndromes.[2] The differential diagnoses of hemihypertrophy are Proteus syndrome, Beckwith–Weidemann syndrome, Silver–Russell syndrome (SRS), Klippel–Trenaunay–Weber syndrome (KTWS), Bannayan–Riley syndrome,

Encephalocraniocutaneous lipomatosis, Hemihyperplasia syndrome, and Neurofibromatosis, Mosaic trisomy 8, Megalencephaly–cutis marmorata telangiectatica and so on.[3,4]

The present case is of hemihypertrophy associated with multiple bilateral giant fibroadenomas and an associated verrucous epidermal nevus but because of absence of other syndromic features and regressive course of the body asymmetry, we postulated it to be IH. IH might be a milder form of syndromes such as KTWS, Beckwith–Wiedemann syndrome, and Proteus syndrome. It has increased risk of childhood cancers similar to Beckwith–Wiedemann syndrome mostly before the age of five years, so screening and follow up should be done for early detection of malignancy until 7 years of age.[1,2] Although fibroadenomas are reported with Beckwith–Wiedemann syndrome, they are not reported with IH.[5] Also, cutaneous and vascular lesions are not associated with nonsyndromic hemihypertrophy.[6] We report this case because of the presence of multiple breast fibroadenomas as well as a cutaneous manifestation (verrucous epidermal nevus) in association with IH.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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