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## Accessory Limb Attached to the Back

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An Accessory limb attached to the back is an unusual abnormality, and it appears that reports are few (1, 2, 3). It is likely, that this condition results from an aborted modified process of twinning. The term "fetus in fetu" is reserved for the malformed masses in which the differentiation of the teratoma is carried to a high degree with the presence of axial differentiation of limbs and organs (4,5). Teratomas are composed of poorly organised tissues derived from each of three layers of the embryonic disc (6,7). It is sometimes difficult to make a clear distinction between teratomas and structures that result from abortive attempts of twinning because of the existency of highly organoid appearance (6).

We here present a 3200 gr full-term boy was born to healthy 2<sup>nd</sup> degree related parents. There was no history of maternal illness, exposure of radiation or drug in take during pergnancy and no history of congenital anomalies in the family. His mother did not come to the antenatal policlinic during her pregnancy. For this reason, this abnormality was determined during delivery. At birth an accessory limb was noted; it resembled a deformed arm and was attached to the back in the midthoracic region. The arm did not have bones, had only the phalangeal bone with two digits (Fig. 1). Deformed accessory arm attached to the back, a rudimentary structure resembling a tiny arm is present in the mid thoracic region was shown in Figure 1. The accessory limb lacked movement, and no pulses were palpable. The child did not have any deformity of hands and feet. There was no evidence of neurological deficit, and on cranial ultrasonography, it was shown that his both lateral ventricles were moderately dilated (20 mm) and there was a bleeding inside both lateral ventricles, meant that he had cranial hemorrhage.



Figure 1. Deformed accessory arm attached to the back in the midthoracic region and neural tube defect in the thoracolumbar region.

Plain x-rays showed that the limb had only the phalangeal bones with two hand digits without the upper arm bones. Accessory limb was present in the

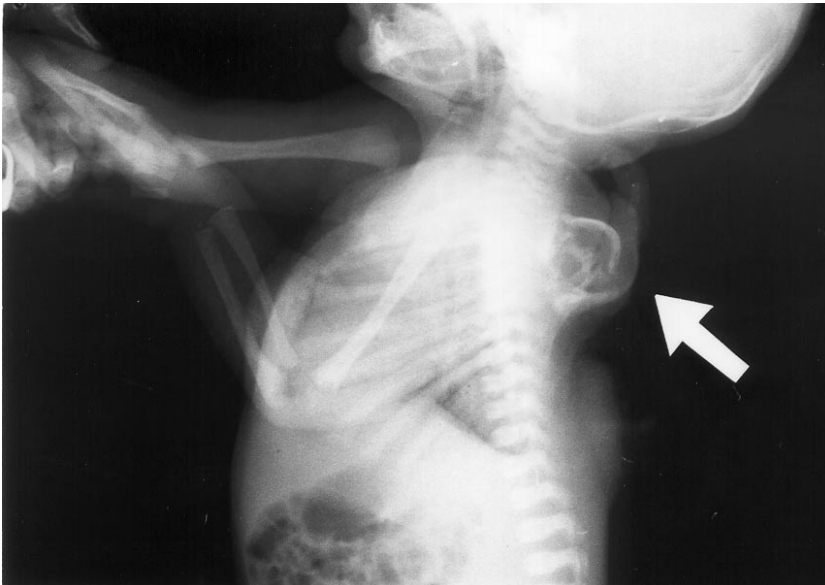


Figure 2. Plain X-ray. The arrow shows that the limb have only phalangeal bones with two hand digits.

midthoracic region (Fig 2). The bony structure of the accessory limb and the evidence of neural tube defect were shown in Fig 2 with lateral radiograph. On pelvic and upper abdominal ultrasonography, there was a cystic mass, approximately 56 mm diameters, that fulfilled pelvis and lying to the level of umbilicus, and this structure was considered as a bladder. His upper abdominal structures were normal.

His chromosomal constitution was normal and there was no abnormality with the pedigree analysis of the family. The case was sporadic. Surgery was decided to be performed when the baby would become 14 days old. But the baby died prior to surgery.

The morphogenesis of this rare malformation is not clear. The limb buds develop from the mesoderm adjacent to the paraxial mesoderm, and a very early splitting of one limb bud may result in this anomaly (2, 8). On the basis of animal experiments, the investigators cited splitting and migration of limb primordia owing to some "mechanical factor" as the etiologic mechanism (8). An accessory scrotum with a rudimentary phallus in one reported case (2) and the presence of an anal dimple in two cases (9) appear to support this hypothesis because these structures are normally located close to the leg attachment to the trunk. When splitting occurs, the embryonic tissue-with the potential to develop into the skin of the scrotum or anal region-could also be carried off and may migrate along with the split limb primordium to the ectopic site; this would explain the presence of these structures when development is completed.

It is also difficult to decide on precise nomenclature for this unusual malformation. On the basis of the presence of a vertebral axis with a normal limb bone arrangement, Chandramouli et al classified their case (of an accessory limb attached to the back) as fetus in fetu (9). Fetus in fetu is distinguished from teratoma by the presence of part or the whole of a vertebral axial skeleton and appropriate arrangement of limbs and other organs with respect to the axis (4). Teratoma is defined a true tumor composed of multiple tissues foreign to those characteristic of the part from which it is derived (6, 7, 10). Previous investigators have hypothesized that fetus in fetu resulted from a modified process of twinning, and have traced a natural progression from normal twins to conjoined symmetrical twins, through parasitic fetuses and fetal inclusion, and finally to teratoma (11). On the other hand, in 1935, Willis opposed this theory and stated that fetus in fetu most likely arises from inclusion of a monozygotic diamniotic twin within the bearer (12). A vertebral axis and appropriate arrangement of other organs or limbs in its relation were the criteria to distinguish a fetus in fetu from a highly differentiated teratoma that arises from very early separation of a focus of multipotential cells.

Cytogenetic studies of the baby showed normal chromosome structure, identical to those of the host (11). In our case, as well as in the other cases reported by Eng et al (11), the chromosome studies of the baby were found normal.

However, such a clear distinction is not always present. A teratoma occurring in association with a fetus

in fetu has been reported (13, 14); however, several reported cases of fetus in fetu did not have a proper vertebral axis (15). Perhaps the cells that split off by a modified process of twinning to produce a parasitic twin, fetus in fetu, teratoma vary in potential for differentiation among different cases and sometimes even within the same patient. The latter could account for the presence of a teratoma in association with a fetus in fetu in the same child (13, 14), and the presence of a teratoma along with a well-developed accessory limb.

Neural tube defect in association with the accessory limb was reported by Krishna et al (2) and was also present in two of three cases reported by Sharma et al (3) (as in our patient). A primary mesodermal defect involving both the limb bud and adjacent paraxial mesoderm may explain the association (2). Another possible explanation is that the migrated mass of

embryonal tissue ultimately forming the accessory limb is present from a very early gestational stage and may cause mechanical interference with closure of the neural tube during later development, which results in spina bifida, or other NTD's.

At the end of this report we still do not know for sure whether this case should be diagnosed as fetus in fetu or teratoma. Perhaps, if we were able to perform magnetic resonance imaging of mediastinum we could make a better distinction for diagnosis.

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