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Pai syndrome: First reported case from Syria

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Abstract

Pai syndrome is a rare congenital disorder consists of a triad of a midline cleft upper lip, cutaneous polyps on the facial midline, and the presence of an intracranial lipoma of the corpus callosum. A 6 days old Syrian male was born at full-term presented by midline cleft lip without cleft palate, cutaneous polyp from the left nostril and onto the cleft lip. Further investigations by CT and MRI of the brain revealed paracallosal lipoma with calcifications. This is the first case of Pai Syndrome to be presented from Syria and the third in the Middle East.

Keywords

Pai syndrome; pericallosal lipoma; midline cleft lip; cutaneous polyp; fetal magnetic resonance imaging; ultrasound; fetus.

Introduction

Pai syndrome is a rare congenital disorder consists of a triad of a midline cleft upper lip, cutaneous polyps on the facial midline, and the presence of an intracranial lipoma of the corpus callosum [1]. This unusual combination was first described by Pai et al. Almost 60 cases were reported in the literature and only few met the full criteria [1,2].

Case Report

We present a case of a 7-day-old boy who presented by his parents to a periphery clinic with a mass protruding from the left nostril and a mass over the upper lip with cleft lip. The case was transferred to Al Razi Surgical Hospital in Aleppo, Syria for further investigation and management (Figure 1).

The patient is the outcome of a non-consanguineous marriage. He was born by a spontaneous vaginal delivery with no complication intrapartum and with a deficient antenatal care due to the low socioeconomic status of the family. He is the first child after three miscarriages. There was no similar case in both parents' family members.

The patient was examined and investigated in Al Razi Surgical Hospital. Examination revealed two separated cutaneous soft masses. First one is polypoidal arise from the left nasal cavity, measuring about 3 cm in length and not causing any breathing difficulty. The other one is also polypoidal mass, soft-skin covered arise from the mid-anterior alveolar process associated with midline cleft lip only. The second mass measures almost 1.5 cm and it is overlayed by the nasal mass. No other orofacial deformities were noted.

Examination of other systems including cardiovascular, neurological, ophthalmological and gastrointestinal were all unremarkable.

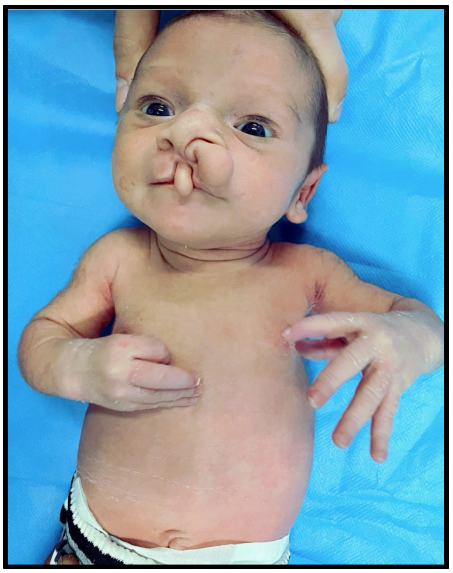

Figure 1: Picture of the newborn showing the nostril polyp and other.

The patient underwent a Computed Topography scan and Magnetic Resonance imaging for further work-up and showed a hypogenesis of the corpus callosum and midline paracallosal lipomas with calcifications (Figure 2).

Both parents were counseled and informed about the finding. Management plan was discussed with them including, a surgical removal of both facial masses beside the cleft lip repair procedure and a referral to Neurology department. Consents were obtained and surgical management was carried on.

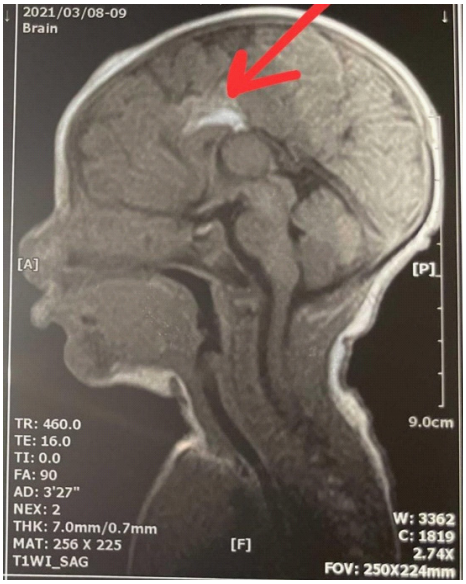

Figure 2: Sagittal section of MRI head with a red arrow showing lipoma of the corpus callosum.

Discussion

Pai et al. described this syndrome in 1987 [1]. Since then, in the literature review, it was rarely to meet the three definitions in one patient. In our case the patient presented with the full criteria of Pai Syndrome (PS) [3]. The three definitions are: Midline facial and nasal skin polyps, midline cleft lip, and lipoma of the corpus callosum [1-3].

The first feature is midline facial and nasal skin polyps, which must present to diagnose PS according to the literature review. The nasal polyp is typically a fibroepithelial polyp (skin tag). In 2007 Castori et al. have noted that this is the most frequent feature [4].

The second feature is midline cleft lip. This anomaly is a challenging deformity and vary in severity [4,5]. The exact etiology of facial clefting is uncertain, the development of midline facial clefts may not be accurate with the classic theory of failure of fusion of normally merging facial processes, though the other theory of an aberration in mesodermal migration appears to be a better explanation [5]. The association of midline cleft lip and nasal anomalies is scarce. Midline clefts were reported in almost 65% of patients with Pai syndrome, with a wide range of severity as mentioned [5].

The third feature is pericallosal lipoma. Generally, Intracranial lipomas are extremely rare lesions, compose almost less than 0.5% of all primary brain tumors [3,4]. It has a good prognosis; mostly asymptomatic, slow-growing lesions and accidentally discovered. There are two types of Lipomas of corpus callosum (LoCCs): anterior (tubulondular) and posterior (curvilinear) [4,5]. In our case it is the first type which is anterior lipoma, known as tubulondular. They are thick, rounded or lobular in shape and may extend into the lateral ventricles. They are usually associated with calcifications with hypogenesis or agenesis of the corpus callosum. Posterior LoCCs are less common, they are thin and elongate along the margin of the corpus callosum which are called curvilinear. These lipomas rarely affect the nervous system, and the neuropsychological development is usually normal in PS. MRI is optimal to diagnose the intracranial lipomas [5,6].

The presentation of the three above mentioned clinical criteria makes our case very unique in nature and up to our knowledge the first to be presented from Middle East. Multidisciplinary team approach with full involvement of the parents in the management of such cases is key factor for optimal outcome and best prognosis.

Declarations

Ethical approval: Given the nature of the article, the management of the patient was not modified by the study, so it was considered exempted from IRB approval.

Patient had an informed consent for the management and permission for using the data for research study if needed.

Authors' contributions: All Authors contributed in literature review, manuscript development and surgical management of the patient.

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