Primary Lymphedema in a Four-Year-Old Boy

Primary lymphedema is usually due to either a congenital absence of, or abnormalities in lymphatic tissue or it is caused by mutations in genes influencing lymphatic development(1). It can be sporadic or hereditary. Primary lymphedema generally presents with a swollen extremity, most often affecting the lower extremities. A four-year-old boy with sporadic type of primary lymphedema is presented here whose symptoms have started at the age of two years with the involvement of the face and both hands and feet.

A four-year-old boy presented with non-pitting edema that involved his hands, feet and the face (Fig. 1). His family noticed the swelling at the age of two years. The amount of edema was described as increasing slowly but never showing regression. There was no family history of such edema. On physical examination, there was non-pitting, non-tender, non-erythematous edema of the hands, feet and the face. The remainder of the physical examination did not reveal pathological finding.

REFERENCES


Fig. 1. Contrast enhanced CT scan of left foot showing the tumor mass.
Complete blood count, sedimentation rate, urinalysis findings, liver function tests including the serum albumin concentration, renal and thyroid function tests were unremarkable. Antinuclear antibodies, anti-nuclear cytoplasmic antibodies, rheumatoid factor, immunoglobulin and C₁ esterase inhibitor levels were also within normal ranges. The chest radiograph and abdominal ultrasound were unremarkable.

Patient’s history, physical examination and laboratory findings suggested primary lymphedema and lymphoscintigraphy revealed hypoplastic lymphatic channels both in the hands and feet.

In the present case, history, physical examination findings and exclusion of other possibilities such as recurrent lymphangitis, neoplasm, surgery or radiation suggested primary lymphedema. The majority of primary cases described in the literature have presented with unilateral lower extremity involvement. Facial and localised symmetrical four extremity involvement has been considered as an uncommon clinical finding(2,3). Interestingly, our patient had symmetrically hand and feet involvement including the wrists and ankles (Fig.1). Swelling of a whole extremity was not observed. He also had face involvement that was more prominent around his eyes and the lips (Fig.1). The case was suggested as sporadic primary lymphedema because there was no family history of such edema.

In conclusion, primary lymphedema in childhood may be sporadic. It can involve the face and four extremities symmetrically. Lymphedema should be considered in children who have progressively increasing edema that is localised, and a lymphoscintigraphic study should be performed in order to confirm the diagnosis.

Fig. 1. Shows facial lymphedema and symmetrical swelling of the hands and feet.
Polio Eradication: Let us Face the Facts and Accept the Reality

In the January 2004 issue of the Bulletin of Polio Eradication Committee of IAP Dr. Jacob John had stated: “it is still possible to see the last case of wild-virus polio in 2003 itself. If that does not happen then it should happen in the first quarter of 2004”. The India Expert Advisory Group concluded at the 26-27 March, 2004 meeting that the transmission of wild polioviruses can be stopped in the country within months. The prophesy turned out to be incorrect as can be seen in Table I.

It would be relevant to state that many polio cases are being missed because of the following two reasons(1): (i) AFP cases where vaccine polioviruses are found in stools are discarded as non-polio. (ii) Wild polioviruses not detected in stool samples of AFP cases. Many such cases are discarded as non-polio even without 60 days follow up.

High incidence of vaccine failure: According to the official data polio incidence in children who had received four or more of doses of OPV was as follows: In year 2000: 58%, in 2001: 60%, in 2002: 44%, and in 2003: 51%. According to Kohler, et al. out of 181 VAPP cases during 1999, 78 children had received five or more doses of OPV before onset of paralysis(2).

High incidence of VAPP: The expected number of VAPP cases every year was 60-75. According to revised data made available by the NPSP the number of VAPP cases were as follows: 1998: 124, 1999: 206, 2000: 151, 2001: 120, 2002: 203. According to my estimates about 300 cases occur every year(1).

It can be said that present eradication program ensures that polio is not eradicated. Polio cases will continue to occur because of vaccine failure and due to mutant vaccine polioviruses. Infected immunocompromised children will continue to spread for prolonged period in the community wild as well as mutant vaccine polioviruses.

It is suggested that following three measures be considered: (i) new guidelines for AFP classification be formulated so that no polio case is missed, (ii) IPV be made available for those children who are immunocompromised or have immunocompromised close contacts, and (iii) the reasons for vaccine failure be determined and appropriate

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