

PPP Syndrome or Ormond's Disease?

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Abstract

The incidence and existence of human immunodeficiency virus concerning women of reproductive age continues to increase globally. The care of HIV-infected women is not simple and must be focused on including the current and future health of these women, the minimization of the risk of maternal-infant transmission and the maintenance of the well-being of the fetus and neonate. Many maternal and obstetrical factors can affect the vertical transmission. The answer to this problem is the optimal medical and obstetrical care.

We have recently described a case of unusual constellation of periophthalmitis, psoriasis, and sinusitis (PPP), which responded to combined corticosteroid and trimethoprim/sulfamethoxazole therapy [1]. Further follow up and additional thoughts conducted us to idea of differential diagnosis of orbital mass (pseudotumor) syndrome. By this way we considered possibility of IGG-4 plasmatic cell infiltration of periophthalmic tissue (Ormond's disease), which was published as a part of retroperitoneal fibrosis and autoimmune pancreatitis and sialoadenitis [2-6]. This syndrome is now known as IGG-4 related systemic disease [7]. For this reason we investigated our patient for blood level of IGG-4, which was of 1130mg/dl compared with normal lower 135 mg/dl. This result was compatible with diagnosis of hyper IGG-4 syndrome. Orbital mass may develop before other manifestations of the Ormond's disease. Everyone should be in suspicion about this plasmatic cell proliferative and infiltrative disease, which may affect every tissue of the body including testis, eyes, salivary glands and cavities. Corticosteroids are drugs of choice and rituximab (anti-CD20) may decline the corticosteroid dependence [8].

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Received August 21, 2014; Accepted October 29, 2014; Published October 31, 2014

Citation: Rozin AP, Toledano K (2014) PPP Syndrome or Ormond's Disease? J Clin Case Rep 4: 437. doi:10.4172/2165-7920.1000437

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