A Case of Lipodystrophy and Polymyositis

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Objective

Describe the first case of polymyositis with lipodystrophy and panniculitis thought to be caused by IgG monoclonal anti-body against IgE.

Introduction

Polymyositis is an immune mediated inflammation of the muscles. Histopathological findings include the endomysial T cells surrounding and invading myofibers, macrophages, myeloid dendritic cells, and plasma cells in association with circulating autoantibodies. Drugs like TNF alpha, interferon alpha and beta, statins and penicillamine are notorious to cause immunologically mediated myopathies.

Lipodystrophy is a condition characterized by partial or complete lack (lipoatrophy) of adipose tissue. There is also apparent accumulation of fat elsewhere in the body. It can be congenital or acquired. Antibodies against adipocyte membrane antigen has been identified in a few cases. Autoimmune disorders such as dermatomyositis, hypothyroidism, pernicious anemia, rheumatoid arthritis, temporal arteritis, or mesangiocapillary glomerulonephritis have been reported in patients with acquired lipodystrophy. It is also seen in patients with HIV, most likely related to antiretroviral therapy.

Panniculitis is inflammation of subcutaneous fat, it is a relatively rare condition, characterized by inflammatory nodules and plaques. Etiology can range from infection, infestation, trauma, enzymatic destruction, malignancy, abnormal deposition and primary or secondary inflammatory conditions.

EMG Summary Table									
	Spontaneous					MUAP			Recruitment
	IA	Fib	PSW	Fasc	H.F.	Amp	Dur.	PPP	Pattern
R. Tibialis anterior	Incr	1+	1+	None	None	N	2-	>50%	Early 2+
R. Gastrocnemius (Medial head)	Incr	1+	1+	None	None	N	1-	N	Early 1+
R. Vastus lateralis	N	None	1+	None	None	N	1-	30-50%	Early 1+
R. First dorsal interosseous	Incr	1+	1+	None	CRDs	N	1-	N	Early 1+
R. Deltoid	Incr	1+	1+	None	None	N	1-	N	Early 1+
R. Thoracic paraspinals (mid)	Incr	1+	1+	None	CRDs	N	2-	30-50%	Early 2+

EMG Summary from Augest 2016

Case Report

This is a case of a 64 year old female with a long standing history of bronchial asthma. In the summer of 2014 she developed a complicated and prolonged course of pneumonia followed by multiple exacerbations of asthma which resulted in frequent ER visits and extended steroid treatment. In approximately 6 months she gained 180 lbs. In Dec 2014 she received Omalizumab as treatment for her asthma and between Dec and Jan received 5 total shots. Soon after she noticed cysts and swelling in her arms, axillae, and thighs, with wasting of her face and arms and loss of subcutaneous fat in her calves. She was diagnosed with partial lipodystrophy. Pathology report from R axillary skin biopsy revealed lobular panniculitis with nodular cystic fat necrosis. Temporally the patient related this to taking Omalizumab.

She was then referred to our Neuromuscular clinic from the Nutrition and Metabolic Disease clinic in 2016 for generalized muscle wasting. She was found to have bilateral distal upper extremity muscle weakness and upper and lower extremity muscle atrophy. Her NCS revealed normal sensory studies and EMG showed evidence of myopathy with membrane instability. MRI (humerus and thigh) confirmed the finding showing intramuscular edema is the deltoid, triceps, supraspinatus and infraspinatus and superficial and deep facial edema in her thigh with reactive lymphadenopathy. Labs revealed a normal Creatinine Kinase and mildly elevated C- reactive protein. The plan was to proceed with IVIG or PLEX based on the results of muscle biopsy however patient was lost to follow up.

She did have an annual follow up at the Nutrition and Metabolic disease clinic where she expressed severe depression due to dysmorphism caused by the lipodystrophy. She was referred to psychiatry and is currently getting fillers for her face for cosmetic reasons.

insert MRIs

Conclusion

Omalizumab is a recombinant, DNA derived, humanized IgG monoclonal antibody and is the only approved biologic designed to target and block IgE. It is indicated for the treatment of allergic asthma and idiopathic chronic urticaria. Its mechanism of action is to reduce IgE, prevent crosslinking of IgE, downregulate IgE receptors (on basophils, mast cells and dendritis cells), limits mast cell degranulation and minimizes release of early and late phase reactants.

As per the safety label and packaging, this medication is contraindicated in severe hypersensitivity reactions. Adverse reactions observed in patients under treatment include arthralgia, arm and leg pain, dizziness, pruritus, dermatitis, ear ache, nasopharyngeal and gastrointestinal infections. Injection site reactions included swelling, erythema, pain, brusing, itching, bleeding and urticarial. There was also an increased risk of cardiovascular and cerebrovascular events reported in patients on treatment with Omalizumab.The warning and precautions in the package insert included anaphylaxis, malignant neoplasms (breast, non melanoma skin, melanoma, prostate, parotid gland), systemic eosinpophillia (churg strauss syndrome), fever, arthritis/arthralgia, rash,lymphadenopathy and high risk for helminthic infection.

Given the singularity of the case it is difficult to infer causality however that being said it is not an impossibility. Other monoclonal antibodies, namely the checkpoint inhibitors have been known to cause Immune related adverse events (IRAEs).Impilimumab and Nivolumab have been reported to have caused polymyositis.To date Omalizumab has not been reported as an offending agent to cause polymyositits or lipodystrophy.

We would suggest to exercise caution while prescribing a biologic agent and weigh pros and cons given novelty of the drug and potential unknown post marketing adverse events.

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