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March 2004

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## Recommended Citation

Khealani, B., Qureshi, R., Wasay, M. (2004). Motor neuropathy associated with Adenocarcinoma of Esophagus. *Journal of Pakistan Medical Association*, 54(3), 165-166.

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# Motor Neuronopathy associated with Adenocarcinoma of Esophagus

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Neurologic paraneoplastic syndromes (NPS) are remote neurologic effects, except metastasis, of systemic cancers. These are a rare group of disorders, commonly associated with small cell lung carcinoma (SCLC).<sup>1</sup> Various NPS have been described, including motor neuronopathy. The motor neuronopathy has commonly been described with small cell carcinoma of lung, breast cancer and lymphoproliferative disorders.<sup>2,3</sup> We report a case of motor neuronopathy in association with large cell adenocarcinoma of esophagus. To our knowledge this is the first reported case of motor neuronopathy associated with adenocarcinoma of the esophagus.

## Case Report

A 57-year old man, smoker, presented with a 2-months history of progressive walking difficulty. He was emaciated and his neurological examination was significant for absent gag reflex, mild proximal limb weakness (power of 4/5 on MRC scale), impaired proprioception and ataxic gait. Chest, abdominal and cardiovascular examination was normal. There was no lymphadenopathy. His complete blood counts, serum electrolytes, liver function tests, CPK were normal. Motor nerve conduction studies of right median, ulnar, posterior tibial and peroneal nerves revealed mildly slow conduction velocities. However, there was no conduction block. Sensory nerve conduction studies of bilateral sural, right median and right ulnar nerves were normal. The EMG findings were suggestive of anterior horn cell disorder (motor neuronopathy). He developed complaints of dysphagia. An upper GI endoscopy revealed a fungating and ulcerated growth at distal esophagus, confirmed on histopathology to be large cell adenocarcinoma. Later patient developed mental status changes and blindness. MRI brain showed multiple hemorrhagic metastasis to bone (sternum). An esophageal stent was placed and patient was managed conservatively.

Later he developed recurrent pneumonia and died two months after the initial presentation.

## Discussion

Neurologic paraneoplastic syndrome was first described by Oppenheim in 1888.<sup>1</sup> The exact incidence of neurologic paraneoplastic syndrome is not known, however incidence in patients with SCLC, the most commonly associated tumor with the syndromes, is less than 3%.<sup>1</sup> The mechanism of development of neurologic paraneoplastic syndrome is not fully understood, however, there is evidence that this is an autoimmune phenomenon.<sup>4</sup> In addition to autoimmunity, role of opportunistic viral infections has been proposed as underlying mechanism for motor neuropathy.<sup>1</sup> The paraneoplastic syndrome can affect any part of nervous system including spinal cord/anterior horn cells. The paraneoplastic motor neuron disease (motor neuronopathy) has commonly been reported in association with SCLC, breast cancer and lymphoproliferative disorders.<sup>2,3</sup> To our knowledge there are only two reports of motor neuronopathy in association with esophageal cancers<sup>5,6</sup> and none of these was adenocarcinoma. We reported motor neuronopathy in association of large cell esophageal adenocarcinoma. EMG findings of our patient are suggestive of anterior horn cell disorder/motor neuronopathy. Our patient did have evidence of reinnervation in three body regions and denervation in only two regions. Furthermore, normal sensory nerve conduction studies, normal CMAP amplitudes, normal distal motor latencies and widespread reinnervating motor unit potentials and patchy active denervation suggest anterior horn cell disorder. Mildly slow conduction velocities could also be seen in axonal neuropathy and multifocal motor neuropathy with conduction block (MMNCB) but preserved compound muscle action potential and absence of conduction block makes these diagnoses unlikely in our

patient. The ataxic gait in our patient could also be a feature of paraneoplastic cerebellar degeneration (PCD). The paraneoplastic motor neuropathy has been described to be part of paraneoplastic encephalomyelopathy/subacute sensory neuronopathy (PEM/SSN) in about 25% of cases.<sup>7</sup> We conclude that our patient has motor neuropathy in setting of esophageal adenocarcinoma, which has not been described previously. This can emphasize the established fact that paraneoplastic syndrome can be initial manifestation of underlying malignancy. This also highlights the importance of electrodiagnostic studies in appropriate settings. We believe that any cancer patient with neurologic symptoms, suggesting peripheral nervous system involvement should undergo nerve conduction and electromyography. Paraneoplastic syndrome can be

associated with adenocarcinoma of esophagus.

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