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polymyositis with 9p+ telomere syndrome]. Here we report a case of polymyositis (PM) with 9p+ telomere syndrome. A 63-year-old female with PM complicated with achalasia was noted to have loose teeth and gingival overgrowth. She also showed progressive hand tremor, proximal muscle weakness and atrophy, and tongue fasciculation. The proximal muscle weakness was aggravated by stress. M-proteins were noted to increase in the patient's serum. The myopathological findings were compatible with a PM diagnosis. Cytogenetic studies using cultured peripheral lymphocytes revealed: (1) hypoploidy of chromosomes 2, 9, 10 and 13, and (2) abnormality of the 9p+ telomere. The chromosome abnormality was more common at the early stage than at the later stage. The present findings indicate that 9p+ telomere syndrome is a genetic disorder having an important role in the pathophysiology of PM.

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