OBJECTIVE: We describe a rare case of primary glioblastoma in the spinal cord and compare the clinical presentation to the few previously described in literature. BACKGROUND: While GBM is the most common primary CNS malignancy in adults, spinal GBM is a very rare condition. Primary intramedullary tumors are infrequent and account for 2-4% of all CNS tumors. Spinal GBM accounts for only 1-5% of all glioblastomas. Moreover, spinal GBM has a predilection for the cervical or thoracic region in >60% of cases. Recognized for its poor prognosis, discussion about those cases, available treatments and outcomes are required to improve survival and ensure quality of life. We described a 49-year-old male, previously healthy, who presented with severe lower back pain for 4 months, associated with paresthesia of the lower extremities. Contrast MRI of the spine revealed a heterogeneously enhancing mass extending from T12-L2. Patient underwent a laminectomy with subtotal resection of intramedullary spinal cord. Histopathology revealed a WHO grade IV tumor. Patient was started on radiotherapy and temozolomide. RESULTS: Surgical pathology revealed WHO grade IV tumor. Patient finished his radiotherapy course and is currently undergoing chemotherapy treatment. CONCLUSION: Intramedullary GBM is a very rare disease. Despite aggressive therapy with surgery, radiation and chemotherapy, all current measures lead to disappointing results, with survival prognosis around 15 months. MRI findings can be unspecific and final diagnosis is made on histologic basis. Excessive resection is difficult in intramedullary lesions, which contributes to the poor prognosis. In addition, leptomeningeal spread is expected in 25-60% of the cases. Most spinal GBM are reported at the cervical level although in the case of our patient, a thoracolumbar level delayed the diagnosis when initially treated as a lower back pain. Early diagnosis, constant discussions and research are required to improve outcome.