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Rhabdomyosarcoma, Immun Thrombocytopenic Purpura and Chronic Kidney Disease Coexistence: A Case Report

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Abstract: Rhabdomyosarcoma (RMS) is a group of soft tissue malignities arises from muscules. In adults head and neck involvement is rare. Immun thrombocytopenic purpura (ITP) is isolated thrombocytopenia with normal results of complete blood count and peripheral blood smear without associated conditions that can cause thrombocytopenia. Autoantibadies against the platelet membrane antigens are thought to be the causative factor. Chronic kidney disease (CKD) is the presence of kidney damage or reduction of glomerular filtration rate more than 3 months. 42 year old woman was diagnosed as craniofacial rhabdomyosarcoma and extended medial maxillectomy was performed. In a routine control, platelet count of 16,000/ mcl was detected 6 months after chemotherapy and ITP was diagnosed. 5 years after the diagnosis of RMS, her urea and creatinine levels began to increase gradually and sustained so she was diagnosed as CKD. In this case RMS, ITP and CKD were diagnosed respectively. During the follow- up period, especially for patients with malignant disorders, physicians should be on alert about additional disorders.
Keywords: Rhabdomyosarcoma, immun thrombocytopenic purpura, chronic kidney disease.

INTRODUCTİON

RMS is a group of soft tissue malignities arises from muscules. In adults head and neck involvement is rare but extremity affliction is more common. The most common localisation of RMS in craniofacial region in adults is ethmoidal sinus with poor prognosis because of the tendency of invading meninges [1-6].

ITP is isolated thrombocytopenia with normal results of complete blood count and peripheral blood smear without associated conditions that can cause thrombocytopenia. Autoantibadies against the platelet membrane antigens are thought to be the causative factor for ITP via increased platelet destruction [7, 8]. CKD is the presence of kidney damage or reduction of glomerular filtration rate more than 3 months. Diabetes and hypertension are the most comon causes of CKD. In end stage CKD dialysis or kidney transplantation are the only therapeutic options [9].

In this case RMS, ITP and CKD were diagnosed respectively. During the follow- up period, especially for patients with malignant disorders, physicians should be on alert about additional disorders.

CASE REPORT

42 year old woman admitted to craniofacial surgery polyclinic because of nasal congestion 10 years ago. In paranasal computerized tomography, in the left maxillary sinus homogenous mass extending to the nasopharynx was detected and extended medial maxillectomy operation was performed. Pathology of the operation material was rhabdomyosarcoma and there were no tumor remnant in the surgical margins. 1 month after surgery, she received radiotherapy for 25 days and 6 cycles of chemotherapy consist of ifosfamide, mesna and doxorubicin. 6 months after the chemotherapy administration in a routine control platelet count of 16,000/ mcl was detected in full blood count. In order to clarify the etiology, direct and indirect Coombs tests, HBsAg, HBeAg, anti-HCV, anti-HIV, brucella tube agglutination, rose bengal tests, ANA, anti-dsDNA, p- ANCA, c- ANCA, immune globulin levels, anti trombin 3, anti- cardiolipin antibodies, protein C and S levels were evaluated and all these tests were negative. In the end of these diagnostic approaches patients diagnosed as chronic immune thrombocytopenic purpura. Patient was treated with 1 mg/ day oral prednisolone, and in the follow up the level of platelet count has risen to 84,000/mcl. Weekly dose of prednisolone was reduced gradually. 5 years after the diagnosis of RMS, her urea and creatinine levels began to increase gradually. Recently her estimated GFR is 20ml/dk and with adequate tension control and avoidance of nephrotoxic agents she does not have uremic complaints.

DISCUSSION

RMS is a heterogenous group of soft tissue malignities arises from muscules and is a member of small round cell tumours. The main histological types are embryonal, alveolar, botryoid and pleomorphic rhabdomyosarcomas. Embryonal type is the most comman subtype and accounts for 70%- 80% of all RMSs. Alveolar subtype is the less frequent subtype and accounts for 10%-20% of all cases. Embryonal histology and sarcoma botryoides have better prognosis compared to alveolar subtype. Although RMS is common in childhood, it is rare in adulthood and accounts for only 2%-5% of all malignant soft tissue tumours with pleomorphic subtype dominantly. In adults head and neck localisation is rare but extremity affliction is relatively common. The most common localisation of RMS in craniofacial region in adults is ethmoidal sinus with poor prognosis because of the tendency to invade meninges [1-6]. RMSs of adults are unusual, predominantly pleomorphic subtype with high-grade cytologic and different biologic characteristics [5, 10-12]. Because of the rarity in adulthood, RMSs are mostly neglected in the differential diagnosis of small round cell tumours and less information is available to guide physicians for the treatment of patients. Especially in craniofacial region other small round cell tumours such as undifferentiated or neuroendocrine carcinoma, lymphoma, olfactory neuroblastoma and melanoma are more common than RMSs in adulthood [13]. Five year survival of head and neck RMSs of adults are poor [14]. Major metastases of RMSs go to lung, lymph nodes and bone marrow [15]. Ifosfamide, adriamycine, etoposide, vincristine and actinomycin are effective therapeutic agents in RMSs [16].

ITP is isolated thrombocytopenia with normal results of complete blood count and peripheral blood smear in a patient without associated conditions that can cause thrombocytopenia such as Acquired Immun Deficiency Syndrome, certain drugs, lymphoprolipherative disorders, myelodysplasia, agammaglobulinemia, systemic lupus erytematosus, alloimmune thrombocytopenia and congenital or hereditary thrombocytopenia. Conditions that can cause blood count and peripheral blood smear abnormalities such as thalassemia minor or iron deficiency does not exclude the ITP diagnosis. Probably autoantibadies against the platelet membrane antigens cause ITP via increased platelet destruction [7, 8]. This disorder is generally chronic, insidous in adults, but one third of the cases are persistent and resistant to treatments. Only 5% of adults with chronic ITP can have spontaneus remission [17]. If platelet counts are low, patients with ITP have the risk of life-threatening bleeding conditions such as intracranial hemorrhage that is the leading cause of the mortalities due to ITP. With the same platelet counts older people have more haemorrhagic complications than younger ones [18, 19]. The diagnosis of ITP is made by confirming the presence of of isolated thrombocytopenia and excluding the other causes of thrombocytopenia with a history, physical examination, complete blood count, peripheral blood smear and sometimes some other necessary tests. The main therapeutic options for ITP are glucocorticoids, intravenous immunoglobulin, intravenous anti-Rho(D),and splenectomy. In refractory cases cyclophosphamide, azathioprine, danazol, vinca alcaloids, ascorbic acid, colchicine, interferon-alpha, combination chemotherapy, epsilon-aminocaproic acid, cyclosporine, protein A immunoadsorbtion, plasma exchange and accessory splenectomy are the other therapeutic options. There is no direct evidence that indicates the reducing effects of these therapeutic interventions on the complications and mortalitiy due to ITP [20].

CKD is the presence of kidney damage or glomerular filtration rate less than 60 ml/min/ 1.73m2 for more than 3 months. Diabetes mellitus, hypertension and primary kidney diseases are the most common causes of CKD all over the world. Although nocturia and fatigue due to anemia are the first signs, with the progression of CKD almost all organ systems are affected by the uremic toxins. Beside the nonspesific treatment such as decreasing the dietary salt consumption, tension and blood glucose control; in advanced stages of CKD renal replacement therapies such as hemodialysis, peritoneal dialysis or kidney transplantation are essential [9].

In this case RMS, ITP and CKD were diagnosed respectively. During the follow- up period, especially for patients with malignant disorders, physicians should be on alert about additional disorders in order not to overlook some crucial conditions.

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