



Some epidemiological characteristics of malignant fibrous histiocytoma in the Province of Vojvodina

Borislava L. Nikolin, Tihomir Dugandžija, Svetlana Salma, Jasna Trifunović, Dragana Vojnović

ABSTRACT

Institute of Oncology Sremska Kamenica, Serbia & Montenegro, Address correspondence to: Dr. Borislava Nikolin, Institute of Oncology Sremska Kamenica, Instituttski put 4, 21204 Sremska Kamenica, Serbia & Montenegro; The manuscript was received: 27.12.2004, Provisionally accepted: 28.12.2004, Accepted for publication: 10.01.2005.

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BACKGROUND: *Malignant fibrous histiocytoma is most common soft tissue sarcoma occurring in late adult life, most in the extremities and in the retroperitoneum, usually developing in deep fascia or skeletal muscle. Because of the aggressive nature of the tumor early and complete surgical removal is indicated. The overall survival rate of patients with malignant fibrous histiocytoma ranges from 36% to 58% at 5 years. Malignant fibrous histiocytoma in Vojvodina is still rare malignancy either in incidence or in mortality in both sexes. The population of Vojvodina is about 2 100 000.*

METHODS: *We used a descriptive epidemiological method to analyze incidence and mortality from malignant fibrous histiocytoma in Vojvodina. This study is based on the Cancer Registry data for the ten-year period from 1993 to 2002.*

RESULTS: *During the observed period 37 cases of malignant fibrous histiocytoma were reported (19 men and 18 women). The average age of patients was 50 years. The body site distribution differed between sexes and malignant fibrous histiocytoma was more common on the trunk (retroperitoneum) in men and on the lower limbs in women. The average annual incidence rate for the observed period was 1.82 per 1 000 000 for both sexes. The incidence rate changed irregularly over the observed period with the peak of 9 cases (4.55 per 1 000 000) in 2001. The overall linear trends of incidence and of mortality showed a slight increasing tendency in both sexes, but not significantly. Concerning mortality, the similar patterns were noticed. The mortality rate was 0.83 per 1 000 000.*

CONCLUSION: *According to epidemiological data we obtained it can be concluded that Province of Vojvodina is not a region with an important risk for malignant fibrous histiocytoma; however, the increase of incidence appearing in both men and women should be taken into account very seriously in future.*

KEY WORDS: *Histiocytoma, Fibrous; Sarcoma; Epidemiology; Mortality; Incidence; Non MeSH Vojvodina*

INTRODUCTION

Malignant fibrous histiocytoma (MFH), described by O'Brien and Stout in 1964, is the most common soft tissue sarcoma occurring in late adult life. Uncertain histogenesis and numerous subtypes make MFH a rather controversial entity. Researchers have postulated both histiocytic and primitive mesenchymal cell theories of origin. In general, the tumor contains both fibroblast like and histiocyte like cells in varying proportions, with spindled and rounded cells exhibiting a storiform arrangement. Five histological subtypes have been described including (1) storiform/pleomorphic (most common), (2) myxoid, (3) giant cell, (4) inflammatory (usually retroperitoneal), and (5) angiomatoid (often located more superficially than other varieties) (1,2).

The clinical stage of the tumor, which is defined by tumor grade, size, and presence of distant metastases, is the most important prognostic factor. Histological subtype and method of surgical treatment are also important prognostic factors. The anatomic site and depth of the primary tumor may also be of prognostic importance but this is controversial. Patients with low-grade, intermediate-grade, and high-grade tumors have 10-year survival rates of

90%, 60%, and 20%, respectively. Patients with tumors smaller than 5 cm at presentation have survival rates of 79%-82%. Patients with tumors of 5-10 cm have survival rates of 62%-68% and those with tumors larger than 10 cm have survival rates of 41%-51%. Distant metastasis most commonly occurs to the lung (90%), bone (8%), and liver (1%). Rate of metastasis varies with the histological subtype from 23% (myxoid) to 50% (giant cell). Positive microscopic margins are associated with decreased disease-free survival while resection with negative microscopic margins decreases the incidence of local recurrence; however, these factors do not have a direct impact on outcome (2-4).

The overall survival rate of patients with MFH ranges from 36%-58% at 5 years; however, patients with retroperitoneal tumors have an overall 5-year survival rate of 15%-20%. MFH occurs more commonly in Caucasian patients than in patients of African or Asian descent. Male-to-female ratio is approximately 2:1. The tumor occurs with a peak incidence in the fifth and sixth decades but an age range of 10 to 90 years is reported. Although the tumor is rare in children, the angiomatoid subtype is the most frequently occurring variety in patients younger than 20 years (4-6). MFH occurs most commonly in the extremities (70%-75%, with lower extremities accounting for 59% of cases), followed by the retroperitoneum.

Tumors typically develop in deep fascia or skeletal muscle. The most common clinical presentation is an enlarging painless soft tissue mass in the thigh, typically 5-10 cm in diameter. Two thirds of tumors are intramuscular. Retroperitoneal MFH usually presents with constitutional symptoms, including fever, malaise, and weight loss. Early and complete surgical removal using wide or radical resection is indicated because of the aggressive nature of the tumor. Biopsy is often necessary to make a diagnosis. Follow-up studies to exclude tumor recurrence usually are performed using MRI (6,7). In order to find out the epidemiological situation concerning MFH in Vojvodina, we made a retrospective analysis of the 10-year period data.

MATERIALS AND METHODS

We used a descriptive epidemiological method to analyze the incidence and mortality from MFH. The data covering a period from 01.01.1993 to 31.12.2002 were obtained from the Cancer Registry of Vojvodina, Institute of oncology Sremska Kamenica (8). The Registry covers the population of the whole territory of Province of Vojvodina with around 2 million inhabitants. The rates were calculated per 1 000 000. The data on cancer reports are routinely collected through the registry network and processed by the registry staff according to the IARC (International Agency for Research on Cancer, Lyon, France) recommendation (9).

RESULTS

MFH in Vojvodina is not among the ten most frequent malignancies neither in incidence nor in mortality in both sexes. During the observed period 37 newly diagnosed cases of MFH were reported, 19 in men and 18 in women, which gives the average annual incidence rate of 1.82 per 1 000 000 for both sexes. The incidence rate changed irregularly over the observed period with the highest peak of 9 cases (4.55 per 1 000 000) in 2001 (Figure 1).

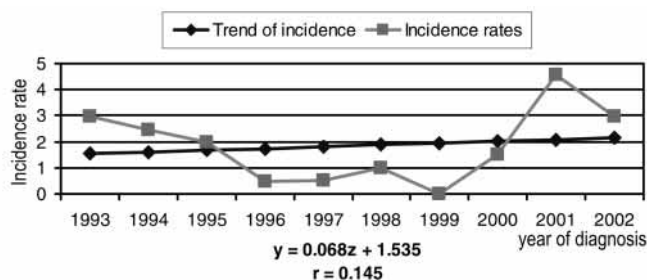


Figure 1. Incidence rates and trend of incidence of MFH in the Province of Vojvodina for the period 1993-2002

The average age of patients was 50 years. The body site distribution differs between sexes and MFH is more common in the trunk (retroperitoneum) in men and in the lower extremities in women. It should be said that the percentage of other and non-specified localization is rather low, which is to be attributed to the high quality of the registration. The overall linear trends of incidence showed a slight increasing tendency (Figure 1).

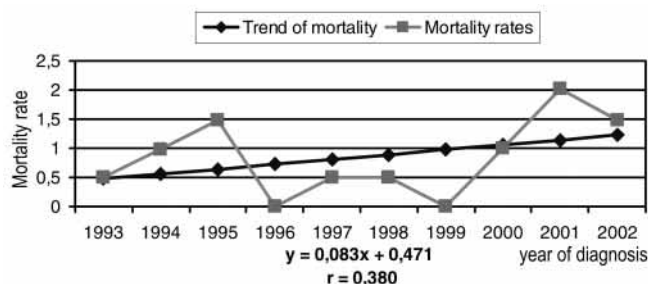


Figure 2. Mortality rates and trend of mortality of MFH in the Province of Vojvodina for the period 1993-2002

Concerning mortality, the similar patterns were noticed. The mortality rate was 0.83/1 000 000 for both sexes. The overall time trend concerning mortality rates through the observed period showed also increasing tendency (Figure 2).

DISCUSSION

The diagnosis of the MFH is based on specific cytomorphological characteristics and immunocyto- and immunohistochemical staining. Literature data show that malignant fibrous histiocytoma is rare disease of histiocytic and/or primitive mesenchymal cell in origin and account for about only 0.025% of all malignancies in adults but it is the most common soft tissue sarcoma occurring in late adult life. MFH is equally disease of men and women. Most frequently, MFH metastasizes in lung, bone and liver (1,10-12). A significant increasing number of cases among both male and female population in the Province of Vojvodina has been registered. The overall linear trend of mortality is increasing. Increased morbidity is followed by increased mortality, which suggests late diagnosis and lower quality of treatment. These results are quite in accordance with the worldwide figures (8,9,13,14).

CONCLUSION

MFH in Vojvodina is still a rare malignancy either in incidence or in mortality in both sexes. The prevention of MFH is impossible because of unknown etiology. By reason of this the early detection is the most important factor for better prognosis. According to epidemiological data we obtained it can be concluded that Province of Vojvodina is not a region with an important risk for malignant fibrous histiocytoma; however, the increase of incidence appearing in both men and women should be taken into account very seriously in future.

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