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Retrospective Study of Managing Group A and B Retinoblastoma

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Abstract

Background: Retinoblastoma is the most common intraocular tumor in children. There are two forms heritable and non-heritable. Leukocoria is the most common presenting sign, accounting for about 56.1% of cases. Strabismus is the second most common mode of presentation. Early retinoblastoma patients (group A and B ICRB) treated by systemic chemotherapy in conjunction with local control have the higher rate of eye salvage.

Aim of Study: To classify retinoblastoma patients presented to Oncology Unit of Ain Shams Department of Ophthalmology to international groups and study presentation, modalities of treatment and treatment outcome of group A and B patients. To see to what extent in Ain Shams Department of Ophthalmology we succeed achieving eye salvage in early retinoblastoma patients group A and B.

Patients and Methods: This study involved patients have been treated at Oncology Unit of Ain Shams Department of Ophthalmology. Data was extracted from recorded files of patients in the period from January 2004 till January 2012. All patients data were subjected to refinement of: Age, sex, date of first visit, complaint, family history, fundus examination, diagnosis, classification to international groups, treatment, follow-up.

Results: Complete tumor regression had occurred in 90% of all group A and B patients following treatment with systemic chemotherapy plus focal laser thermotherapy. Enucleation was needed in 6.7% of all patients. Group A only patients had a 90% percentage of complete tumor regression, with only one patient died from pneumonia (one out of 30). Group B only patients had a 90.5% percentage of complete tumor regression, with only two patients had needed enucleation (2 out of 30). EBR was needed in 6.7% of all patients. Age at presentation mean was 1.09 year with range (0.1-3). Regarding presenting symptoms, 80% of patients presented with leukocoria and 13.3% with squint. Family history was negative in 86.7% and positive in 13.3%.

Conclusion: Our conclusion is that early retinoblastoma patients (group A and B ICRB) treated by systemic chemotherapy in conjunction with local control have resulted in ocular salvage rates exceeding 90% for early intraocular disease.

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Key Words: *Retinoblastoma – International classification for intraocular retinoblastoma (ICRB) – Chemoreduction – Laser thermotherapy.*

Introduction

RETINOBLASTOMA is a rare form of cancer that rapidly develops from the immature cells of a retina, the light-detecting tissue of the eye. It is the most common malignant cancer of the eye in children. Though most children survive this cancer, they may lose their vision in the affected eye(s) or need to have the eye removed **[1,2]**.

About 1 out of 3 retinoblastomas is caused by a mutation in the retinoblastoma 1 (RB1) gene (long arm of chromosome 13 band 13q14) that is present in all the cells of the child's body. But of these cases, only about 1 in 4 is inherited from one of the child's parents. In the rest, the gene mutation is not inherited, but occurs during early development in the womb. Most of the remaining 2 out of 3 retinoblastomas occur as a result of a random RB 1 gene mutation that occurs only in one cell of one eye **[3]**.

Intraocularly, it exhibits a variety of growth patterns, endophytic growth occurs when the tumor breaks through the internal limiting membrane and has an ophthalmic appearance of a white-to-cream mass showing either no surface vessels or small irregular tumor vessels, exophytic growth occurs in the subretinal space and is often associated with subretinal fluid accumulation and retinal detachment, diffuse infiltrating growth is a rare subtype comprising 1.5% of all retinoblastomas characterized by a relatively flat infiltration of the retina by tumor cells but without a discrete tumor mass **[4,5]**.

Leukocoria (white pupillary reflex or cat's eye reflex) is the most common presenting sign, accounting for about 56.1% of cases. Strabismus,

which occurs as a result of visual loss, is the second most common mode of presentation. Thus, funduscopic examination through a well-dilated pupil must be performed in all cases of childhood strabismus. Other less common signs and symptoms of retinoblastoma include: Vision problems, red painful eye with glaucoma, hyphema, bulging of the eye, Anisocoria, Heterochromia iridis, Orbital cellulitis & Nystagmus **[6]**.

Patients noted to have signs of retinoblastoma should undergo complete eye examination including an estimation of the patient's visual acuity for both eyes. A dilated fundus examination with indirect ophthalmoscopy should be completed since ancillary diagnostic studies play only a secondary role when the fundus can be visualized clearly. Imaging tests (Imaging tests use X-rays, sound waves, magnetic fields, or radioactive substances to create pictures of the inside of the body) will be done to help confirm it and to find out how far it may have spread within the eye and possibly to other parts of the body **[7]**.

The outlook (prognosis) for children with retinoblastoma depends, to some extent, on the cancer's stage. The stage is also an important factor in choosing treatment. The International Classification for Intraocular Retinoblastoma (ICRB) is the newer retinoblastoma staging system. In this staging system, intraocular retinoblastoma is differentiated into 5 groups, from A to E. A indicates a better prognosis, and E indicates a poorer prognosis using existing treatment modalities. According to the ICRB, patients within groups A, B, and C had a significant chance of ocular salvage and avoidance of EBRT while those within group D had a much higher rate of treatment failure, with approximately one half requiring External Beam Radiation Therapy (EBRT) or enucleation. Group E eyes were intended to be managed with primary enucleation **[8-10]**.

Prior to the International Classification for Intraocular Retinoblastoma, the Reese-Ellsworth classification system was the most useful system when (EBRT) was the standard of treatment for eye salvage. However, now that chemotherapy has supplanted radiation, this classification system is not as predictive of outcome and survival **[8,9]**.

The goals of treatment for retinoblastoma are to preserve the patient's life and salvage the affected globe, to preserve as much vision as possible and to limit the risk of second cancers later in life, which can be caused by treatment, particularly in children with hereditary retinoblastoma **[11,12]**.

Overall, more than 9 in 10 children with retinoblastoma are cured. The chances of long-term survival are much better if the tumor has not spread outside the eye **[13]**.

The main types of treatment for retinoblastoma are chemotherapy, thermotherapy (using a type of laser to apply heat to kill small tumors), photocoagulation (using lasers to kill small tumors or the blood vessels that feed them), Cryotherapy (using cold to freeze and kill small tumors), radiation therapy and surgery (enucleation).

Sometimes more than one type of treatment may be used. The treatment options are based on the extent of the cancer and other factors **[14]**.

Patients with treated retinoblastoma should be monitored with examination under anesthesia every 3-4 months until age 3-4 years, after which they are examined under anesthesia every 6 months until age 5-6 years and then annually thereafter. At about age 8 years, most patients can tolerate a dilated fundus examination in the office without anesthesia and can be examined annually in the office thereafter **[15,16]**.

In the developed world, retinoblastoma has one of the best cure rates of all childhood cancers (95-98%), with more than nine out of every ten sufferers surviving into adulthood. In the UK, around 40 to 50 new cases are diagnosed each year. Good prognosis depends upon early presentation of the child in health facility. Late presentation of the child in hospital is associated with poor prognosis **[17-19]**.

Aim of the work:

To classify retinoblastoma patients presented to Oncology Unit of Ain Shams Department of Ophthalmology to international groups and study presentation, modalities of treatment and treatment outcome of group A and B patients. To see to what extent in Ain Shams Department of Ophthalmology we succeed achieving eye salvage in early retinoblastoma patients group A and B.

Patients and Methods

Type of study: Retrospective observational study.

Study setting: This study involved patients have been treated at Oncology Unit of Ain Shams Department of Ophthalmology.

Study period: Data was extracted from recorded files of patients in the period from January 2004 till January 2012.

Study population:

Inclusion criteria: Data extracted from files of all Retinoblastoma patients presented to Oncology Unit of Ain Shams Department of Ophthalmology during study period.

Exclusion criteria: Incomplete patients' data. Examination details not appropriate enough for classification. Lost follow-up. Patients of other groups than A and B.

Sampling method: Convenience sampling.

Sample size: 30 eyes of 30Pts.

Ethical considerations: Confidentiality of records were considered.

Study procedure: All patients data were subjected to refinement of: Age, sex, date of first visit, complaint, family history, fundus examination, diagnosis, classification to international groups, treatment, follow-up.

Details of dilated fundus examination using cyclopentolate with indirect ophthalmoscopy with scleral indentation documented in patients' files such size and number of tumor and other associated pathology (assessment of the mass color, location, overlying retinal detachment and subretinal fluid, presence of vitreous seeds (focal or diffuse) and determination of its class and subretinal seeds).

Classification to international groups:

The outlook (prognosis) for children with retinoblastoma depends, to some extent, on the cancer's stage. The stage is also an important factor in choosing treatment. The International Classification for Intraocular Retinoblastoma is the newer retinoblastoma staging system. In this staging system, intraocular retinoblastoma is differentiated into 5 groups, from A to E. A indicates a better prognosis, and E indicates a poorer prognosis using existing treatment modalities.

In our study we had 9 group (A) patients and 21 group (B) patients.

Tumor size was 3mm or less and was located at least 3mm from the foveola and 1.5mm from OD in all group (A) patients. In group (B) patients, tumor size was 3mm or less in 4Pts. and more than 3mm in 17Pts. Tumor was located at least 3mm from the foveola and 1.5mm from OD in 8Pts. and near OD or foveola in 13Pts.

Treatment: Primary line treatment was collected from documented data.

For group (A), focal diode laser transpupillary thermotherapy was performed until complete tumor regression plus systemic chemotherapy if needed.

For group (B), systemic chemotherapy ± focal diode laser transpupillary thermotherapy as needed. Other lines of treatment (external beam radiation-Enucleation) was needed only in group (B) in case of primary treatment failure.

Chemotherapy treatment protocol: A six-treatment cycles of chemoreduction therapy with vincristine (1.5mg/m^2), etoposide (200mg/m^2), and carboplatin (560mg/m^2) was administered and a complete blood profile was obtained at monthly intervals. Eyes that responded to chemoreduction therapy received focal transpupillary thermotherapy.

Thermotherapy was used for retinoblastomas of 3mm or less in base diameter and 3mm or less in thickness without vitreous seeds located at or posterior to the equator. Occasionally, fish flesh portions of larger diameter retinoblastomas that regressed in a type III manner were treated with thermotherapy using an IRIDEX OcuLight SLx (California, USA) Infrared (810nm) laser system. Treatments were administered in the form of monthly cycles, with chemotherapy administered on the first 2-3 days of every month. The laser was set at a continuous mode with a spot diameter ranging from 0.5 to 1.2mm and the power was adjusted at 300-400mW and was applied for 1-5min per spot till covering 100% of tumor area, the end point was a gentle, light gray color change within the tumor without causing vascular spasm or rapid tumor whitening.

Non regressed tumors were treated with additional chemoreduction and focal treatment. When these methods failed, EBRT (a dose ranging from 35.0 to 45.0Gy) was administered.

Follow-up visits:

The patients were followed-up under anesthesia by fundus examination every 4 weeks during treatment period until complete response.

After complete response, follow-up carried every 3 months was extracted from files for at least 1 year.

Statistical analysis:

Data were collected, revised, coded and entered to the Statistical Package for Social Science (IBM SPSS) version 23. The quantitative data were presented as mean, standard deviations and ranges when parametric. Also, qualitative variables were presented as number and percentages. The compar-

ison between groups regarding qualitative data was done by using Chi-square test. The comparison between two independent groups regarding quantitative data with parametric distribution was done by using Independent *t*-test. The confidence interval was set to 95% and the margin of error accepted was set to 5%. So, the p-value was considered significant as the following: $p>0.05$: Non significant (NS). $p<0.05$: Significant (S). $p<0.01$: Highly significant (HS).

Results

Table (1): Clinical characteristics of group (A).

		Group A (No.=9)
Age:	Mean ± SD	0.88±0.55 year
	Range	0.25-1.8 year
Sex:	Males	6 (66.7%)
	Females	3 (33.3%)
Complaint:	Brother of RB	1 (11.1%)
	Leucocoria	5 (55.6%)
	Squint	3 (33.3%)
Family history:	Negative	7 (77.8%)
	Positive	2 (22.2%)

This table shows that age of presentation of retinoblastoma patients in group (A) patients was at range of 3 to 21 months, with average age of 10.56 months with standard deviation ±6.6 months. Percentage of positive family history among group (A) was 22.2%, 2 of 9 group (A) patients.

Table (2): Group (A) fully dilated fundus examination details.

		Group A (No.=9)
Tumor size:	≤3mm	9 (100.0%)
	>3mm	0 (0.0%)
Near OD of foveola:	No	9 (100.0%)
	Yes	0 (0.0%)

This table shows that tumor size was 3mm or less and was located at least 3mm from the foveola and 1.5mm from OD in all group (A) patients.

Table (3): First line treatment options of group (A) patients.

		Group A (No.=9)
Chemotherapy:	No	2 (22.2%)
	Yes	7 (77.8%)
Chemotherapy cycles:	Mean ± SD	7.86±2.85
	Range	6-12
Laser:	No	0 (0.0%)
	Yes	9 (100.0%)
Laser sessions:	Mean ± SD	6.44±2.35
	Range	3-12

These table and figure show that 77.8% of group (A) Pts. had received systemic chemotherapy (7 out of 9) and 100% underwent laser thermotherapy. Average count of chemotherapy cycles was 7.86 with SD ±2.85 (range 6-12). Average number of laser sessions was 6 with SD ±2.35 (range 3-12).

Table (4): Primary outcome of group (A).

		Group A (No.=9)	
Regression:	No	0	0.0%
	Yes	9	100.0%
Continue chemotherapy:	No	9	100.0%
	Yes	0	0.0%
Add laser sessions:	No	9	100.0%
	Yes	0	0.0%
EBR:	No	9	100.0%
	Yes	0	0.0%
Enucleation:	No	9	100.0%
	Yes	0	0.0%

These table and figure show that 100% of group (A) Pts. Showed tumor regression (9 out of 9). There was no need to continue chemotherapy or add laser sessions or EBR or enucleation.

Table (5): Final outcome of group (A).

	Group A (No.=9)
Observation time or period (months):	
Mean ± SD	30.11±7.74 months
Range	15-41 months
Final outcome:	
Complete regression	8 (88.9%)
Enucleation	0 (0.0%)
Dead	1 (11.1%)

This table shows that average period of observation of group (A) patients was 30.11 months with SD ±7.74. Complete repression had been achieved in 90% of Pts. (8 out of 9). One Pt. died of pneumonia (no available data if it was related to chemotherapy or not) during follow-up.

Table (6): Clinical characteristics of group (B).

		Group B (No.=21)
Age:	Mean ± SD	1.17±0.74 year
	Range	0.1-3 year
Sex:	Males	11 (52.38%)
	Females	10 (47.62%)
Complaint:	Leucocoria	19 (90.5%)
	Mass	1 (4.8%)
	Squint	1 (4.8%)
Family history:	Negative	19 (90.5%)
	Positive	2 (9.5%)

This table shows that age of presentation of retinoblastoma patients in group (B) patients was at range of 1 month to 3 years, with average age of 1.17 year with standard deviation ±0.74 year. Percentage of positive family history among group (B) was 9.5%, 2 of 21 group (B) patients.

Table (7): Group (B) fully dilated fundus examination details.

		Group B (No.=21)
Tumor size:	≤3mm	4 (19.0%)
	>3mm	17 (81.0%)
Near OD of foveola:	No	8 (38.1%)
	Yes	13 (61.9%)

This table shows that tumor size was 3mm or less in 4Pt. (19%) and more than 3mm in 17Pt. (81%). Tumor was located at least 3mm from the foveola and 1.5mm from OD in 8Pt. (38.1%) and near OD or foveola in 13Pt. (61.9%).

Table (8): First line treatment options of group (B) patients.

		Group B (No.=21)
Chemotherapy:	No	1 (4.8%)
	Yes	20 (95.2%)
Chemotherapy cycles:	Mean ± SD	7.95±2.24
	Range	6-12
Laser:	No	5 (23.8%)
	Yes	16 (76.2%)
Laser sessions:	Mean ± SD	5.75±0.68
	Range	4-6

These table and figure show that 95.2% of group (B) Pts. had received systemic chemotherapy (20 out of 21) and 76.2% underwent laser thermotherapy. Average count of chemotherapy cycles was 7.95 with SD ±2.24 (range 6-12). Average number of laser sessions was 5.75 with SD ±0.68 (range 4-6).

Table (9): Primary outcome of group (B).

		Group A	
		No.	%
Regression:	No	3	14.3%
	Yes	18	85.7%
Continue chemotherapy:	No	19	90.5%
	Yes	2	9.5%
Add laser sessions:	No	20	95.2%
	Yes	1	4.8%
EBR:	No	19	90.5%
	Yes	2	9.5%
Enucleation:	No	19	90.5%
	Yes	2	9.5%

These table and figure show that 85.7% of group (B) Pts. Showed tumor regression (18 out of 21) and 14.3% showed no primary response (3 out of 21). Chemotherapy cycles was continued in 9.5% of Pts. (2 out of 21). Laser sessions was added in 4.8% of Pts. (1 out of 21). EBR was needed in 9.5% of Pts. (2 out of 21). Enucleation also was needed in 9.5% of Pts. (2 out of 21).

Table (10): Final outcome of group (B).

	Group B (No.=21)
Observation time or period (months):	
Mean ± SD	32.43±6.55 months
Range	24-55 months
Final outcome:	
Complete regression	19 (90.5%)
Enucleation	2 (9.5%)

This table shows that average period of observation of group (B) patients was 32.43 months with SD ±6.55. Complete regression had been achieved in 90.5% of Pts. (19 out of 21). 9.5% of cases (2 out of 21) ended with enucleation.

Table (11): Follow-up and final outcome comparison between group (A) and group (B) of our patients.

	Group A No.=9	Group B No.=21	Test value	p-value	Sig.
Observation time or period (months):					
Mean ± SD	30.11±7.74	32.43±6.55	–0.842•	0.407	NS
Range	15-41	24-55			
Final outcome:					
Complete regression	8 (88.9%)	19 (90.5%)	3.192*	0.203	NS
Encleation	0 (0.0%)	2 (9.5%)			
Dead	1 (11.1%)	0 (0.0%)			

p-value >0.05: Non significant.
p-value <0.05: Significant.
p-value <0.01: Highly significant.
*: Chi-square test.
•: Independent t-test.

This table shows that no significant difference had been found in observation period nor final outcome between group (A) and group (B) patients. 88.9% of group (A) patients (8 out of 9) had com-

plete regression and only one patient 11.1% (1 out of 9) died from pneumonia. 90.5% of group (B) patients (19 out of 21) had complete regression and only two patients 9.5% (2 out of 21) had eye enucleated.

Discussion

Managing early retinoblastoma (group A and B ICRB) has the importance that we can achieve eye salvage with the recent favorable easy line of treatment consisting of chemoreduction mainly with or without focal therapy.

The ICRB (international classification of intraocular retinoblastoma) was designed to simplify retinoblastoma classification and to predict treatment success with current methods, specifically CRD (chemoreduction). This classification was not intended to predict life prognosis or visual outcome. It was intended to predict globe outcome, specifically, avoidance probability of enucleation and EBRT after CRD. In this study, we have noticed that patients within groups A and B had a considerable chance for globe salvage and avoidance of EBRT **[8-10]**.

According to our study: Complete tumor regression had occurred in 90% of all group A and B patients following treatment with systemic chemotherapy plus focal laser thermotherapy. Enucleation was needed in 6.7% of all patients. Group A only patients had a 90% percentage of complete tumor regression, with only one patient died from pneumonia (one out of 30). Group B only patients had a 90.5% percentage of complete tumor regression, with only two patients had needed enucleation (2 out of 30).

EBR was needed in 6.7% of all patients. Age at presentation mean was 1.09 year with range (0.1-3). Regarding presenting symptoms, 80% of patients presented with leukocoria and 13.3% with squint. Family history was negative in 86.7% and positive in 13.3%.

So our study results support the findings from previous studies on retinoblastoma; Shields et al. (2008), found the success of chemoreduction protocol in 100% of 23 group A patients and 93% of 96 group B patients.

Novetsky **[10]**, found 16% enucleation percentage in 10 group A patients +53 group B patients who received single-agent intravenous carboplatin. Age range was (0.03-2.5 year).

Friedman **[20]**, found that systemic neoadjuvant chemotherapy (Vincristine and Carboplatin (VC), accompanied by local ophthalmic therapies such as cryotherapy, diode laser thermotherapy, or brachytherapy, as required) for 21 Group B intraocular retinoblastoma patients achieved eye salvage percentage of 85%, enucleation percentage was 15% and EBR percentage was 10%.

Lumbroso **[21]**, describe the efficacy of conservative management of retinoblastoma by an association of conservative ocular therapies and chemothermotherapy (carboplatin) and found that eye salvage was 100% in 16 group A patients, 94.59% in 37 group B patients and EBR followed by enucleation was needed in 5.41% of group B patients.

Friedman **[22]**, found that chemoreduction (vincristine, etoposide, and Carboplatin) with local ophthalmic therapy for early intraocular retinoblastoma (Reese-Ellsworth groups 1 and 2) resulted in avoidance of EBR and Enucleation in 100% of 18 patients.

In comparison with our study:

Zage **[2]**, found that the vision salvage rate without EBRT for 22 eyes with ICRB groups A and B tumors treated with chemoreduction (carboplatin and etoposide) plus focal retinal therapy was 77.3%, 100% for 7 group A patients alone and 66.7% for 15 group B patients alone. Enucleation was needed in 5 out of 15 group B patients. The mean age at diagnosis was 0.975 year (range 0.1-5.3 year).

Factors that may have influenced our results and conclusions:

Filing system, all our data retrieved from patients records, so any inaccurate details could change results specially the data of tumor size and location as it disrupts group classification of patients in study based on international classification of intraocular retinoblastoma like our study.

Study period, our study results depended on at least one-year period of non-progressed tumor, more post-treatment follow-up time could lead to more assured results of re-progressed tumors.

Conclusion:

Our conclusion is that early retinoblastoma patients (group A and B ICRB) treated by systemic chemotherapy in conjunction with local control have resulted in ocular salvage rates exceeding 90% for early intraocular disease.

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دراسة إرتجاعية لمناظرة مجموعة (آ) و (ب) من الورم الآرومى الشبكى

الورم الآرومى الشبكى هو الورم الأولى الأكثر شيوعاً فى العين فى الطفولة، تبيض الحدقة (المنعكس الحد قى الأبيض آو منعكس عين القط) هو العرض الأكثر شيوعاً، وهو ما يمثل حوالى ٥٦.١٪ من الحالات. الحول، والذى يحدث نتيجة لفقد العين للرؤية، هو ثانى أكثر عرض شيوعاً. الأنواع الرئيسية لعلاج الورم الآرومى الشبكى هى العلاج الكيميائى، التخثير الضوئى، العلاج الحرارى، العلاج بالتبريد، العملية الجراحية والعلاج الإشعاعى وفى بعض الآحيان يمكن إستخدام أكثر من نوع واحد من العلاج. تم تصميم (التصنيف الدولى للورم الآرومى الشبكى داخل العين) لتبسيط تصنيف ورم الآرومة الشبكى والتنبؤ بنجاح العلاج مع الآساليب الحالية، وتحديداً (المعالجة الكيميائية). الغرض من هذا التصنيف هو التنبؤ بنتيجة كرة العين، وتحديداً، إحتمالية تجنب إزالة كرة العين آو تعريض المريض للإشعاع بعد العلاج الكيميائى. فى هذه الدراسة، لاحظنا آن المرضى فى المجموعات آ و ب لديهم فرصة كبيرة لإنقاذ كرة العين وتجنب العلاج الإشعاعى.

قد قمنا بعمل دراسة إرتجاعية على مرضى الورم الآرومى الشبكى الذين توافدوا على وحدة الآورام فى قسم طب وجراحة العيون بمستشفى جامعة عين شمس. وكان الهدف من الدراسة تصنيف مرضى الورم الآرومى الشبكى المترددين على وحدة الآورام فى قسم عين شمس لطب وجراحة العيون طبقاً لمجموعات التصنيف الدولى ودراسة الآعراض السريرية وطرق العلاج المختلفة ونتائج العلاج لمجموعتى (آ) و (ب). ولمعرفة لآى حد قد نجحنا فى قسم الرمد بجامعة عين شمس فى إنقاذ العين لمرضى الورم الآرومى الشبكى الحديث مجموعة (آ) و (ب). وقد خلصت النتائج إلى آنه حدث تحوف كامل للورم فى ٩٠٪ من جميع مرضى المجموعة (آ) و (ب) بعد العلاج الكيميائى المجموعى بالإضافة إلى العلاج بالليزر الحرارى البؤرى، كان هناك حاجة إلى إزالة العين فى ٦.٧٪ من جميع المرضى، كان لدى المجموعة (آ) فقط نسبة ٩٠٪ من الإنحدار الكامل للورم، مع وفاة مريض واحد فقط بسبب الإلتهاب الرئوى (واحد من ٣٠) ، كان للمرضى فى المجموعة (ب) فقط نسبة ٩٠.٥ من الإنحدار الكامل للورم، مع وجود مريضين فقط بحاجة إلى إستئصال (٢ من ٣٠)، كانت هناك حاجة لعلاج إشعاعى فى ٦.٧٪ من جميع المرضى، متوسط العمر كان ١.٠٩ سنة مع النطاق (١.٠-٣)، فيما يتعلق بالآعراض، قدم ٨٠٪ من المرضى يعانون من تبيض الحدقة و١٣.٣٪ من الحول وكان تاريخ العائلة سلبياً فى ٨٦.٧ وإيجابياً فى ١٣.٣٪.