Linda's Story – Epitheliod Hemangioendothelioma (EHE) of the Aorta

I have a very rare cancer, which was extremely difficult to diagnose.

In January 2002 I had a pain in my left side, which got progressively worse. In March I saw my GP, who said she thought the pain was muscular. I saw a different Dr, about the same pain in June and another one in September. Both of these Dr's also said they thought the pain was muscular.

I asked the third Dr what I could do about the pain and he referred me to a physiotherapist, who gave me some exercises to do. She said she thought my problem was caused by poor deportment and suggested I get some McKenzie back supports. I took her advice and purchased one for my office chair and another for my car. The first physiotherapist went on maternity leave and her successor said because I was waking up during the night in pain; it could be that I needed a new bed. (I did purchase one – it made no difference).

After being discharged from the physiotherapist in November I paid privately to see an Osteopath. I had several treatments, which unfortunately did not help the pain.

I went back to the Dr's surgery in December 2002 and saw a fourth Dr who sent me for a chest x-ray. During the follow up appointment I explained that I had now been in pain for a year and asked to be referred to a Consultant at the Cheltenham General Hospital.

I had to wait until April 2003. The Consultant suggested various tests, including an ECG, blood test, urine test, x-ray, bone scan and an ultra sound scan. When I saw the Consultant in July he explained that nothing had shown up in any of the tests and would see me in 6 months time. I said that I was very disappointed because by this time I was in a great deal of pain, which was worsening. I did ask for either a CAT or MRI scan, saying if it was a question of cost I was willing to pay myself. He offered to refer me to a pain clinic, but I explained that I wanted to find out the cause of the pain, not how to manage it.

I was generally in good health, other than the pain. I felt that the Consultant thought I was wasting his time because none of the tests showed up any problems.

In late July I saw my Dr, and told her that I was unhappy that no one had been able to find the cause of my pain, which was almost unbearable. I had not been able to sleep throughout the night for about 7 months and felt exhausted. She said that the Consultant had actually referred me to a chest specialist.

In August I saw a chest specialist. He organised a CAT scan; this took place in September. I got the results in October which showed that I had a tumour wrapped right around my aorta. He explained that I would need to have surgery to remove the tumour. I waited a very fraught 6 weeks for the operation.

On the day I was due to be admitted I was advised to ring in before actually travelling to the hospital. When I did so, was advised that there was no bed available for me. I rang the Surgeon's secretary who promised to see what she could do to help. She called back at around 1600 hours to say she had tried unsuccessfully to find me a bed. I was devastated. However, at 1930 I had a call from the hospital to say that a bed had become available and did I want it. My husband drove me the 75 miles to the hospital arriving at 2100.

I had the operation in December at the Heartlands Hospital in Birmingham; and was discharged within 6 days. The Cardiothoracic Surgeon explained he was unable to remove the tumour because of where it was sited (around the aorta); he had only been able to take biopsies. He also found growths on my left lung and took biopsies of those too.

In February I saw one of the Registrars who explained that the lumps on the lung were also the same as the tumour around the aorta. They were malignant and diagnosed as paragangliomas. I was informed that because the cancer had spread it was now deemed to be terminal as the condition was incurable. The next step was to see an Oncologist.

After surgery I suffered a lot of sickness and acid reflux and lost 3 stones in weight. My Dr said I looked 'haunted'. I felt so ill and was in a lot of pain.

I had a CAT scan in March (11 weeks after my operation) to see if the tumours had grown since my operation. The results showed it to be a slow growing tumour. That afternoon I was injected with an isotope solution. For the next 3 days I had scans to see if the tumours had taken up the isotopes; unfortunately this did not work.

After the last scan in early March I was scheduled to see the Palliative Care Dr. She was concerned because I was in absolute agony and coughing up blood. I had difficulty in breathing and the pain was as if someone had taken a knife to me. I was admitted to hospital straight away for tests, which showed I had 2 blood clots on my left lung. I was treated with Heparin and Warfarin. After this time the pain was much easier to manage. I was hospitalised for 8 days.

At the end of March I saw the Oncologist who said that he wanted to treat me with a new drug called Imatinib (Glivec), however, I would need to have a PET scan before and after taking the drugs to check for suitability. The cost of each scan was £1,000; he would need to check with the local Primary Care Trust (PCT) that funding was available for the scans. Because the cancer was rare he wasn't sure if the PCT would fund the scans. Fortunately, after much debate, they did.

In April I was given the cytotoxic drug called Imatinib. This drug is to try to stop the tumours from growing. Further CT scans have shown that the tumours I had are slow growing and had not grown very much.

Because the primary tumour was so rare I asked to have a second opinion. This was carried out at The Royal Marsden Hospital in London. The outcome was that the hospital confirmed that surgery to remove the tumour was not possible. The pathology department said the tumours were in fact, Epitheliod Haemangioendotheliomas, not paragangliomas as first thought.

This kind of tumour can occur at any age and affects equal numbers of men and women. It is not known why some people develop this cancer and there are no known risk factors. In most cases the tumour is found in a blood vessel such as a vein or artery, but it may also begin in soft tissue.

Often Epitheliod Haemangioendotheliomas are made up of cells that are inactive and so the tumour grows very slowly. However, sometimes the tumours are made up of cells that are more active (atypical or malignant). These cells look different when examined under the microscope and may be more likely to spread to other sites of the body (mestastasize).

Treatment depends on a number of factors such as the location of the tumour, the speed at which the tumour is growing and its appearance under a microscope (grade) and whether it has spread to other parts of the body (stage). A person's general health and symptoms are also taken into account.

Where possible, treatment is with surgery to remove the tumour. However, this is not always an option if blood vessels are affected, particularly if these are larger blood vessels (as in my case). Other treatments that may be considered include radiotherapy and chemotherapy.

In July I was referred to a Consultant Radiologist, who advised against radiotherapy, as it could damage surrounding tissue and perhaps make it difficult for me to eat or drink.

After doing a lot of research on the Internet, I contacted the Cabrini Clinic in New York in April 2004, regarding a treatment called fractionated stereotactic body radiosurgery. At that time the Consultant had about 11 years of experience using this method of treatment.
In fact, the name 'radiosurgery' is a misnomer. Body Radiosurgery is pinpoint precision radiation using multiple, finely-contoured beams from many different angles - all directed at the cancer minimizing radiation to normal healthy tissue while the patient's body is maintained in a stable, reproducible position.
The attractiveness of this non-invasive technique is that is can be used when surgery, standard radiation and chemotherapy are not viable options or have not produced the desired results.
Body Radiosurgery can be implemented in those patients who desire a non-invasive treatment alternative that has great effectiveness in selected situations. Because Body Radiosurgery is so precise, higher than normal doses of radiation can be given over a shorter period of time. Since higher radiation doses can be given, fewer treatments are necessary compared to standard external beam radiation, yet results are superior. In fact, recent analysis shows Body Radiosurgery is superior to direct isotope infusion into organs such as the pancreas.
It is easy to see the attractiveness of normal tissue protection. Since normal healthy structures are shielded, higher doses of radiation can be delivered to the cancer with anticipation of greater success.
On 1st November 2004, I started treatment in New York. After the consultation I had a body fitting, this involved lying on a black plastic bag which had been filled with resin and catalyst chemicals. I laid on this for about 15 minutes and it set around my body. This mould was then placed on a stretcher, which had measurement marks down each side. I was also given small tattoos - one on each leg and two on my chest. I then had a CT scan while resting on the mould on the stretcher.

The multi-disciplinary 'team' looked at all of my notes, pathology slides, various scans including the one I had there and decided on a course of treatment for me. This involved 5 sessions to treat the main tumour around my aorta and 3 sessions to treat the two tumours in my left lung. The treatment was carried out over 3 weeks and took place at 1930 hours each time. Each treatment involved being placed at the same place on the stretcher (for absolute accuracy) and then the actual radiation session was about 2 minutes. I did not suffer from any side effects whatsoever.

The NHS does use this kind of treatment; however, at present it is only used on brain, head and neck tumours. Because this treatment was not available to me in the UK and not 'recognised' by the NHS, I had to pay for it myself. The total expenses for the trip for the treatment, flights, taxis, accommodation, etc., were around £12,000. It was fortunate that I had an endowment policy which matured so instead of using it to pay the mortgage; I used it to pay for my treatment.

There was no guarantee that the treatment would work - however, they have an 85% success rate, so I felt it was worth the risk and was very optimistic. Since then I have felt so much better; my eating has been much easier and I have put on all the weight I lost (and more!).

In February 2005 I had another CT scan. This showed that the tumours had not grown. I was in much less pain. Instead of taking pain killers every 4 – 5 hours, I was taking them once a day. Although still not sleeping throughout the night, I felt so much better in myself.

Further CT scans have shown that the main tumour has reduced in size by a third and the small tumours on my lung and liver have remained stable.

Each time I have a scan I purchase a copy to send to Dr Lederman in New York. He and his multidisciplinary

team look at the scan and give me a second opinion. This has been carried out without my being charged for this service.

I'm still in some discomfort and get 'sweats' throughout the day and night. However, these have reduced since starting on acupuncture therapy.

I am sure that my survival to date has been strengthened by the fractionated stereotactic body radiosurgery I had in New York.

The Oncology department in Cheltenham General Hospital treats patients from Gloucestershire, Herefordshire and South Worcestershire and comes under the 3 Counties Cancer Network. I now chair a Cancer User's Group for Gloucestershire and am a member of a Cancer Partnership Group in an effort to make positive suggestions to the NHS on how cancer services can be improved.

Should any people who are suffering from cancer want to talk about the treatment I had in New York, I would be very happy to hear from them.

The website for Dr Lederman is: www.RSNY.org His e-mail address is: gil.lederman@rsny.org

The website for this extremely rare type of cancer (EHE) is www.cancerssociety.com/forum/viewmessages.cfm?forum=36&topic=108